

- Cardiology (290)
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2600 MCQ with Illustrated Answer
Full Topics Note
14 Chapter

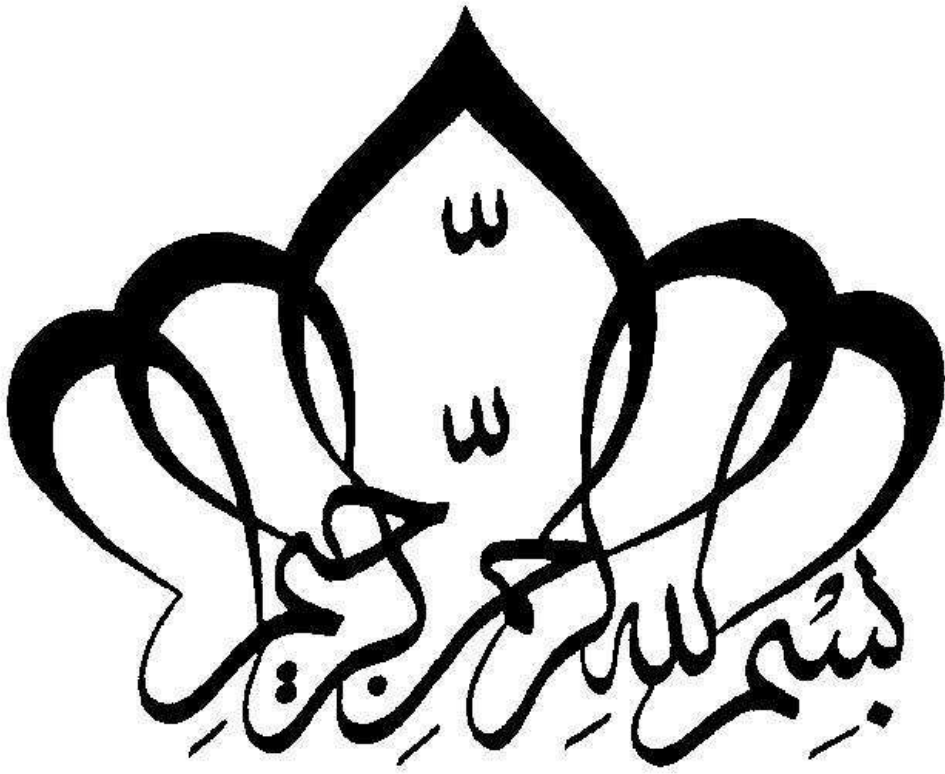
Neurology

P. M MCQ Bank 2017



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قَالُوا سُبْحَانَكَ لَا عِلْمَ لَنَا إِلَّا مَا عَلَّمْتَنَا
۞ إِنَّكَ أَنْتَ الْعَلِيمُ الْحَكِيمُ

إهداء

للكل آية من كتاب الله حفظناها ولكل معلومة قرأناها ونسيناها لعل الله يحمينا من النسيان ويروها إلينا
للكل شخص وعونا له في ظهر الغيب لعل الله أن يستجيب دعائنا
للكل مريض قرأ له ما أله لعل الله يشفيه ويعير إليه صحته وعافيته
للكل لحظة تألم بها أحببنا لعل الله لا يريهم الألم ولا يضييهم مرة أخرى
للكل موقف اجتجت به عيناى فلم تلبيانى لعل الله لا يضرنى بهما في حياتى
للكل حالم صاحب أهراف ومباوى لعل الله يحقق له مقاصده حتى يرضيه ويرضى عنه
للكل مجتهد لعل الله يجعل له من التوفيق أوفر حظ ونصيب ويهون له الأسباب كلها من حيث لا يدري من غير ضراء مضرة ولا فتنة مضلة
للكل من فقر شيئا في الحياة لعل الله يعيره إليه أو يعوضه بما يسعده
للكل شخص ابتعدنا عنه لعل الله يجمعه بمن هم أفضل منا
وأخيرا وليس آخرا لأهلنا لعل الله يجعلها لهم صرقة جارية ترو إليهم قليلا من فضلهم علينا وتكون سببا يلم شملنا تحت سقف واحد بعد أن
هجرتنا الحروب والمصائب

يقول أحد الصالحين

إِذَا رَأَيْتَ أَنَّ اللَّهَ وَفَقَكَ لِإِخْوَةِ تَعْيْنِكَ عَلَى الْخَيْرِ.. فَأَعْلَمْ أَنَّ اللَّهَ يَرِيدُ بِكَ خَيْرًا !! وَإِذَا وَفَقَكَ اللَّهَ لِلدُّعَاءِ.. فَأَعْلَمْ أَنَّهُ يَرِيدُ أَنْ يُعْطِيَكَ !! وَإِذَا
وَفَقَكَ لِقَضَاءِ حَوَائِجِ النَّاسِ فَأَعْلَمْ بِأَنَّهُ لَنْ يَتَخَلَّى عَنْكَ !! وَإِذَا وَفَقَكَ اللَّهَ لِذِكْرِهِ.. فَأَعْلَمْ أَنَّهُ يُحِبُّكَ !!.. وَإِذَا أُحِبَّكَ أَحَدٌ.. وَنَصَرَكَ.. وَأَيَّرَكَ..
وَأُسْتَجَابَ لِدُعَائِكَ !!

This issue of **MRCPPART1 MADE EASY 2017** contains the following:

- Full topic note.
- Around 2600 MCQs from **PM 2017 BANK** covering 14 chapter in internal medicine
- External links for the related guidelines (such as NICE, SIGN, RCP ...etc).
- Comments with illustrative pictures and links to external media.
- Most Recommended QBank for MRCP Part 1, Arab Board, Jordanian Board and other internal medicine assessment exams.

Please Note that

Each chapter begins with notes section. These notes are repeated under the Questions in the website. In these collections the notes are not repeated under each question except in some to avoid unnecessary repetition. For clarity and convenience the title of the related note is only mentioned after the comments in case the reader want to refer back to the respective note in the notes section.

The illustration below is to show and Example

☐	Minimal change disease
☐	Post-streptococcal glomerulonephritis
☐	Goodpasture's syndrome

Goodpasture's syndrome

- IgG deposits on renal biopsy
- anti-GBM antibodies

• These changes are characteristic of Goodpasture's syndrome.

[Discuss and give feedback](#)

Goodpasture's syndrome

Comment of the Q

title of the note in case the reader want to refer back to the notes sections

MRCPPART1 MADE EASY 2017

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إعداد وتنسيق

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&

A. MURAD

لا تنسونا من صالح دعائكم

ما كان من خطأ فمن أنفسنا وما كان من توفيق فمن الله

تم بحمد الله وتوفيقه ومنه

جعل الله عملنا متقبلاً خالصاً لوجهه الكريم

Reference ranges

Reference ranges vary according to individual labs. All values are for adults unless otherwise stated

Full blood count

Haemoglobin	Men: 13.5-18 g/dl Women: 11.5-16 g/dl
Mean cell volume	82-100 fl
Platelets	150-400 * 10 ⁹ /l
White blood cells	4-11 * 10 ⁹ /l

Urea and electrolytes

Sodium	135-145 mmol/l
Potassium	3.5 - 5.0 mmol/l
Urea	2.0-7 mmol/l
Creatinine	55-120 umol/l
Bicarbonate	22-28 mmol/l
Chloride	95-105 mmol/l

Liver function tests

Bilirubin	3-17 umol/l
Alanine transferase (ALT)	3-40 iu/l
Aspartate transaminase (AST)	3-30 iu/l
Alkaline phosphatase (ALP)	30-100 umol/l
Gamma glutamyl transferase (yGT)	8-60 u/l
Albumin	35-50 g/l
Total protein	60-80 g/l

Other haematology

Erythrocyte sedimentation rate (ESR)	Men: < (age / 2) mm/hr Women: < ((age + 10) / 2) mm/hr
Prothrombin time (PT)	10-14 secs
Activated partial thromboplastin time (APTT)	25-35 secs
Ferritin	20-230 ng/ml
Vitamin B12	200-900 ng/l
Folate	3.0 nmol/l
Reticulocytes	0.5-1.5%
D-Dimer	< 400 ng/ml

Other biochemistry

Calcium	2.1-2.6 mmol/l
Phosphate	0.8-1.4 mmol/l
CRP	< 10 mg/l
Thyroid stimulating hormone (TSH)	0.5-5.5 mu/l
Free thyroxine (T4)	9-18 pmol/l
Total thyroxine (T4)	70-140 nmol/l
Amylase	70-300 u/l

Uric acid	0.18-0.48 mmol/l
-----------	------------------

Arterial blood gases

pH	7.35 - 7.45
pCO2	4.5 - 6.0 kPa
pO2	10 - 14 kPa

Lipids

Desirable lipid values depend on other risk factors for cardiovascular disease, below is just a guide:

Total cholesterol	< 5 mmol/l
Triglycerides	< 2 mmol/l
HDL cholesterol	> 1 mmol/l
LDL cholesterol	< 3 mmol/l

Absence seizures	Notes
Absent ankle jerks, extensor plantars	Notes
Acoustic neuroma	Notes
Acute confusional state	Notes
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Absence seizures

Absence seizures (petit mal) are a form of generalised epilepsy that is mostly seen in children. The typical age of onset of 3-10 years old and girls are affected twice as commonly as boys

Features

- absences last a few seconds and are associated with a quick recovery
- seizures may be provoked by hyperventilation or stress
- the child is usually unaware of the seizure
- they may occur many times a day
- EEG: bilateral, symmetrical 3Hz spike and wave pattern

Management

- sodium valproate and ethosuximide are first-line treatment
- good prognosis - 90-95% become seizure free in adolescence

External Links:

[NICE](#)

2012 Epilepsy guidelines

Absent ankle jerks, extensor plantars

Typically caused by lesion producing both upper motor neuron (extensor plantars) and lower motor neuron (absent ankle jerk) signs

Causes:

- subacute combined degeneration of the cord
 - motor neuron disease
 - Friedreich's ataxia
 - syringomyelia
 - taboparesis (syphilis)
 - conus medullaris lesion
-

Acoustic neuroma

Acoustic neuromas (more correctly called vestibular schwannomas) account for approximately five percent of intracranial tumours and 90 percent of cerebellopontine angle

Features can be predicted by the affected cranial nerves:

- cranial nerve VIII: hearing loss, vertigo, tinnitus
- cranial nerve V: absent corneal reflex
- cranial nerve VII: facial palsy

Bilateral acoustic neuromas are seen in neurofibromatosis type 2

MRI of the cerebellopontine angle is the investigation of choice

Acute confusional state

Acute confusional state is also known as delirium or acute organic brain syndrome. It affects up to 30% of elderly patients admitted to hospital.

Features - wide variety of presentations

- memory disturbances (loss of short term > long term)
- may be very agitated or withdrawn
- disorientation
- mood change
- visual hallucinations
- disturbed sleep cycle
- poor attention

Management

- treatment of underlying cause
 - modification of environment
 - the 2006 Royal College of Physicians publication 'The prevention, diagnosis and management of delirium in older people: concise guidelines' recommended haloperidol 0.5 mg as the first-line sedative
 - the 2010 NICE delirium guidelines advocate the use of haloperidol or olanzapine
-

External Links:

[NICE](#)

2010 Delirium guidelines

[RCP](#)

Delirium guidelines

Alzheimer's disease

Alzheimer's disease is a progressive degenerative disease of the brain accounting for the majority of dementia seen in the UK

Genetics

- most cases are sporadic
- 5% of cases are inherited as an autosomal dominant trait
- mutations in the amyloid precursor protein (chromosome 21), presenilin 1 (chromosome 14) and presenilin 2 (chromosome 1) genes are thought to cause the inherited form
- apolipoprotein E allele E4 - encodes a cholesterol transport protein

Pathological changes

- macroscopic: widespread cerebral atrophy, particularly involving the cortex and hippocampus
- microscopic: cortical plaques due to deposition of type A-Beta-amyloid protein and intraneuronal neurofibrillary tangles caused by abnormal aggregation of the tau protein
- biochemical: there is a deficit of acetylcholine from damage to an ascending forebrain projection

Neurofibrillary tangles

- paired helical filaments are partly made from a protein called tau
- in AD tau proteins are excessively phosphorylated.

Management

- NICE now recommend the three acetylcholinesterase inhibitors (donepezil, galantamine and rivastigmine) as options for managing mild to moderate Alzheimer's disease
- memantine (a NMDA receptor antagonist) is reserved for patients with moderate - severe Alzheimer's

External Links:

[NICE](#)

2011 Dementia guidelines

Aphasia

The table below lists the major types of aphasia. Remember that dysarthria is different and refers to a motor speech disorder.

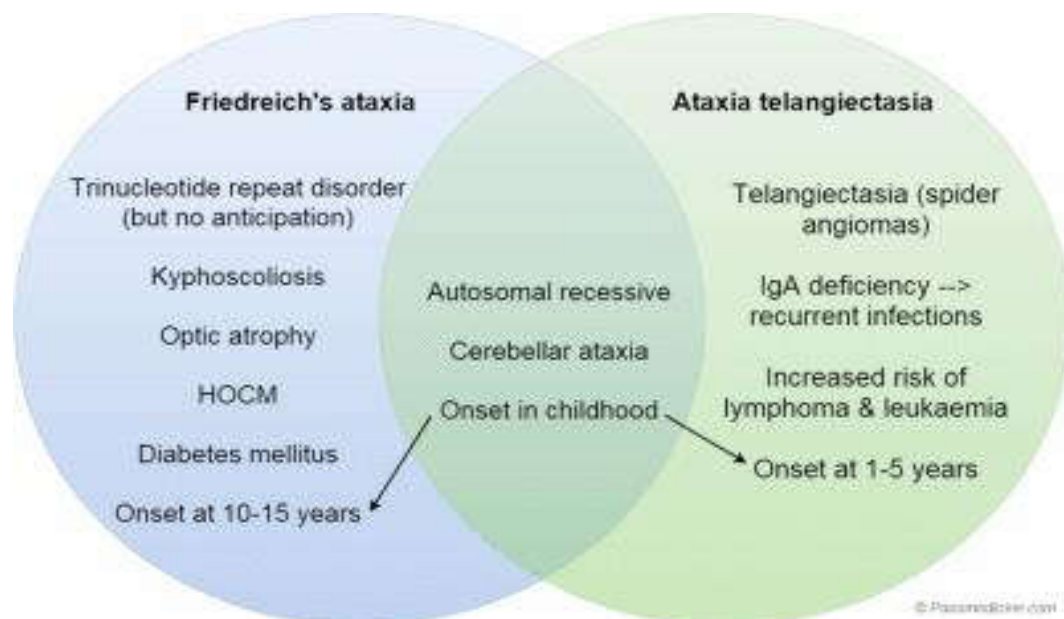
Type of aphasia	Notes
Wernicke's (receptive) aphasia	Due to a lesion of the superior temporal gyrus This area 'forms' the speech before 'sending it' to Broca's area. Lesions result in sentences that make no sense, word substitution and neologisms but speech remains fluent Comprehension is impaired
Broca's (expressive) aphasia	Due to a lesion of the inferior frontal gyrus Speech is non-fluent, laboured, and halting Comprehension is normal
Conduction aphasia	Classically due to a stroke affecting the arcuate fasciculus - the connection between Wernicke's and Broca's area Speech is fluent but repetition is poor. Aware of the errors they are making Comprehension is normal
Global aphasia	Large lesion affecting all 3 of the above areas resulting in severe expressive and receptive aphasia

Ataxic telangiectasia

Ataxic telangiectasia is an autosomal recessive disorder caused by a defect in the ATM gene which encodes for DNA repair enzymes. It is one of the inherited combined immunodeficiency disorders. It typically presents in early childhood with abnormal movements.

Features

- cerebellar ataxia
- telangiectasia (spider angiomas)
- IgA deficiency resulting in recurrent chest infections
- 10% risk of developing malignancy, lymphoma or leukaemia, but also non-lymphoid tumours



Comparison of Friedreich's ataxia and ataxic telangiectasia. Note in particular how ataxic telangiectasia tends to present much earlier, often at the age of 1-2 years

Atrial septal defects

Atrial septal defects (ASDs) are the most likely congenital heart defect to be found in adulthood. They carry a significant mortality, with 50% of patients being dead at 50 years. Two types of ASDs are recognised, ostium secundum and ostium primum. Ostium secundum are the most common

Features

- ejection systolic murmur, fixed splitting of S2
- embolism may pass from venous system to left side of heart causing a stroke

Ostium secundum (70% of ASDs)

- associated with Holt-Oram syndrome (tri-phalangeal thumbs)
- ECG: RBBB with RAD

Ostium primum

- present earlier than ostium secundum defects
- associated with abnormal AV valves
- ECG: RBBB with LAD, prolonged PR interval

Autonomic neuropathy

Features

- impotence, inability to sweat, postural hypotension
- postural hypotension e.g. drop of 30/15 mmHg
- loss of decrease in heart rate following deep breathing
- pupils: dilates following adrenaline instillation.

Causes

- diabetes
- Guillain-Barre syndrome
- multisystem atrophy (MSA), Shy-Drager syndrome
- Parkinson's
- infections: HIV, Chagas' disease, neurosyphilis
- drugs: antihypertensives, tricyclics
- craniopharyngioma

Bell's palsy

Bell's palsy may be defined as an acute, unilateral, idiopathic, facial nerve paralysis. The aetiology is unknown although the role of the herpes simplex virus has been investigated previously. The peak incidence is 20-40 years and the condition is more common in pregnant women.

Features

- lower motor neuron facial nerve palsy - forehead affected*
- patients may also notice post-auricular pain (may precede paralysis), altered taste, dry eyes, hyperacusis

Management

- in the past a variety of treatment options have been proposed including no treatment, prednisolone only and a combination of aciclovir and prednisolone
- following a National Institute for Health randomised controlled trial it is now recommended that prednisolone 1mg/kg for 10 days should be prescribed for patients within 72 hours of onset of Bell's palsy. Adding in aciclovir gives no additional benefit
- eye care is important - prescription of artificial tears and eye lubricants should be considered

Prognosis

- if untreated around 15% of patients have permanent moderate to severe weakness

*upper motor neuron lesion 'spares' upper face

External Links:

[Clinical Knowledge Summaries](#)

Bell's palsy guidelines

Benign paroxysmal positional vertigo

◆ Benign paroxysmal positional vertigo (BPPV) is one of the most common causes of vertigo encountered. It is characterised by the sudden onset of dizziness and vertigo triggered by changes in head position. The average age of onset is 55 years and it is less common in younger patients.

Features

- vertigo triggered by change in head position (e.g. rolling over in bed or gazing upwards)
- may be associated with nausea
- each episode typically lasts 10-20 seconds
- positive Dix-Hallpike manoeuvre

BPPV has a good prognosis and usually resolves spontaneously after a few weeks to months. Symptomatic relief may be gained by:

- Epley manoeuvre (successful in around 80% of cases)
- teaching the patient exercises they can do themselves at home, for example Brandt-Daroff exercises

Medication is often prescribed (e.g. Betahistine) but it tends to be of limited value.

External Links:

[Clinical Knowledge Summaries](#)

Benign paroxysmal positional vertigo guidelines

[YouTube](#)

Hallpike Test and Epley Maneuver

Brain lesions

The following neurological disorders/features may allow localisation of a brain lesion:
Gross anatomy

Parietal lobe lesions:

- sensory inattention
- apraxias
- astereognosis (tactile agnosia)
- inferior homonymous quadrantanopia
- Gerstmann's syndrome (lesion of dominant parietal): alexia, acalculia, finger agnosia and right-left disorientation

Occipital lobe lesions:

- homonymous hemianopia (with macula sparing)
- cortical blindness
- visual agnosia

Temporal lobe lesion:

- Wernicke's aphasia: this area 'forms' the speech before 'sending it' to Brocas area. Lesions result in word substitution, neologisms but speech remains fluent
- superior homonymous quadrantanopia
- auditory agnosia
- prosopagnosia (difficulty recognising faces)

Frontal lobes lesions:

- expressive (Broca's) aphasia: located on the posterior aspect of the frontal lobe, in the inferior frontal gyrus. Speech is non-fluent, laboured, and halting
- disinhibition
- perseveration
- anosmia
- inability to generate a list.

Cerebellum lesions:

- midline lesions: gait and truncal ataxia
- hemisphere lesions: intention tremor, past pointing, dysidiadokinesis, nystagmus.

More specific areas

Area	Associated conditions
Medial thalamus and mammillary bodies of the hypothalamus	Wernicke and Korsakoff syndrome
Subthalamic nucleus of the basal ganglia	Hemiballism
Striatum (caudate nucleus) of the basal ganglia	Huntington chorea
Substantia nigra of the basal ganglia	Parkinson's disease
Amygdala	Kluver-Bucy syndrome (hypersexuality, hyperorality, hyperphagia, visual agnosia)

External Links:

[American Speech-Language-Hearing Association](#)

Common Classifications of Aphasia

Carbamazepine

♦Carbamazepine is chemically similar to the tricyclic antidepressant drugs. It is most commonly used in the treatment of epilepsy, particularly partial seizures, where carbamazepine remains a first-line medication. **Other uses include:**

- neuropathic pain (e.g. trigeminal neuralgia, diabetic neuropathy)
- bipolar disorder

Mechanism of action

- binds to sodium channels increases their refractory period

Adverse effects:

- P450 enzyme inducer
- dizziness and ataxia
- drowsiness
- headache
- visual disturbances (especially diplopia)
- Steven-Johnson syndrome
- leucopenia and agranulocytosis
- syndrome of inappropriate ADH secretion

Cataplexy

Cataplexy describes the sudden and transient loss of muscular tone caused by strong emotion (e.g. laughter, being frightened). Around two-thirds of patients with narcolepsy have cataplexy.

Features range from buckling knees to collapse.

Cerebellar syndrome

Unilateral cerebellar lesions cause ipsilateral signs.

Causes:

- Friedreich's ataxia, ataxic telangiectasia
 - neoplastic: cerebellar haemangioma
 - stroke
 - alcohol
 - multiple sclerosis
 - hypothyroidism
 - drugs: phenytoin, lead poisoning
 - paraneoplastic e.g. secondary to lung cancer
-

Cerebrospinal fluid: raised lymphocytes

Normal values of cerebrospinal fluid (CSF) are as follows:

- pressure = 60-150 mm (patient recumbent)
- protein = 0.2-0.4 g/l
- glucose = $> 2/3$ blood glucose
- cells: red cells = 0, white cells $< 5/\text{mm}^3$.

The following conditions are associated with raised lymphocytes:

- viral meningitis/encephalitis
 - TB meningitis
 - partially treated bacterial meningitis
 - Lyme disease
 - Behcet's, SLE
 - lymphoma, leukaemia
-

Cerebrospinal fluid: raised protein

Normal values of cerebrospinal fluid (CSF) are as follows:

- pressure = 60-150 mm (patient recumbent)
- protein = 0.2-0.4 g/l
- glucose = $> 2/3$ blood glucose
- cells: red cells = 0, white cells $< 5/\text{mm}^3$

The following conditions are associated with raised protein levels:

- Guillain-Barre syndrome
- tuberculous, fungal and bacterial meningitis
- spinal block (Froin's syndrome*)
- viral encephalitis

*describes an increase in CSF protein below a spinal canal blockage (e.g. tumour, disc, infection)

Chorea

Chorea describes involuntary, rapid, jerky movements which often move from one part of the body to another. Slower, sinuous movement of the limbs is termed athetosis. Chorea is caused by damage to the basal ganglia, especially the caudate nucleus.

Causes of chorea

- Huntington's disease, Wilson's disease, ataxic telangiectasia
- SLE, anti-phospholipid syndrome
- rheumatic fever: Sydenham's chorea
- drugs: oral contraceptive pill, L-dopa, antipsychotics
- neuroacanthocytosis
- pregnancy: chorea gravidarum
- thyrotoxicosis
- polycythaemia rubra vera
- carbon monoxide poisoning
- cerebrovascular disease

External Links:

[Postgraduate Medical Journal](#)

Review of chorea

Cluster headache

Cluster headaches* are more common in men (5:1) and smokers.

Features:

- pain typical occurs once or twice a day, each episode lasting 15 mins - 2 hours
- clusters typically last 4-12 weeks
- intense pain around one eye (recurrent attacks 'always' affect same side)
- patient is restless during an attack
- accompanied by redness, lacrimation, lid swelling
- nasal stuffiness
- miosis and ptosis in a minority

Management:

- acute: 100% oxygen, subcutaneous or a nasal triptan
- prophylaxis: verapamil, prednisolone
- NICE recommend seeking specialist advice from a neurologist if a patient develops cluster headaches with respect to neuroimaging

*some neurologists use the term trigeminal autonomic cephalgia to group a number of conditions including cluster headache, paroxysmal hemicrania and short-lived unilateral neuralgiform headache with conjunctival injection and tearing (SUNCT). It is recommended such patients are referred for specialist assessment as specific treatment may be required, for example it is known paroxysmal hemicrania responds very well to indomethacin

External Links:

[NICE](#)

2012 Headache guideline

[SIGN](#)

2008 Diagnosis and management of headache in adults

Cranial nerves

The table below lists the major characteristics of the 12 cranial nerves:

Nerve	Functions	Clinical	Pathway/foramen
I (Olfactory)	Smell		Cribriform plate
II (Optic)	Sight		Optic canal
III (Oculomotor)	Eye movement (MR, IO, SR, IR) Pupil constriction Accommodation Eyelid opening	Palsy results in • ptosis • 'down and out' eye • dilated, fixed pupil	Superior orbital fissure (SOF)
IV (Trochlear)	Eye movement (SO)	Palsy results in defective downward gaze → vertical diplopia	SOF
V (Trigeminal)	Facial sensation Mastication	Lesions may cause: • trigeminal neuralgia • loss of corneal reflex (afferent) • loss of facial sensation • paralysis of mastication muscles • deviation of jaw to weak side	V ₁ : SOF, V ₂ : Foramen rotundum, V ₃ : Foramen ovale
VI (Abducens)	Eye movement (LR)	Palsy results in defective abduction → horizontal diplopia	SOF
VII (Facial)	Facial movement Taste (anterior 2/3rds of tongue) Lacrimation Salivation	Lesions may result in: • flaccid paralysis of upper + lower face • loss of corneal reflex (efferent) • loss of taste • hyperacusis	Internal auditory meatus
VIII (Vestibulocochlear)	Hearing, balance	Hearing loss Vertigo, nystagmus Acoustic neuromas are Schwann cell tumours of the cochlear nerve	Internal auditory meatus

IX (Glossopharyngeal)	Taste (posterior 1/3rd of tongue) Salivation Swallowing Mediates input from carotid body & sinus	Lesions may result in; - hypersensitive carotid sinus reflex - loss of gag reflex (afferent)	Jugular foramen
X (Vagus)	Phonation Swallowing Innervates viscera	Lesions may result in; - uvula deviates away from site of lesion - loss of gag reflex (efferent)	Jugular foramen
XI (Accessory)	Head and shoulder movement	Lesions may result in; weakness turning head to contralateral side	Jugular foramen
XII (Hypoglossal)	Tongue movement	Tongue deviates towards side of lesion	Hypoglossal canal

Some cranial nerves are motor, some sensory and some are both. The most useful mnemonic is given below.

CN I ----->XII

Some Say **M**arry **M**oney **B**ut **M**y **B**rother Says **B**ig **B**rain **M**atter **M**ost

S = Sensory, **M** = Motor, **B** = Both

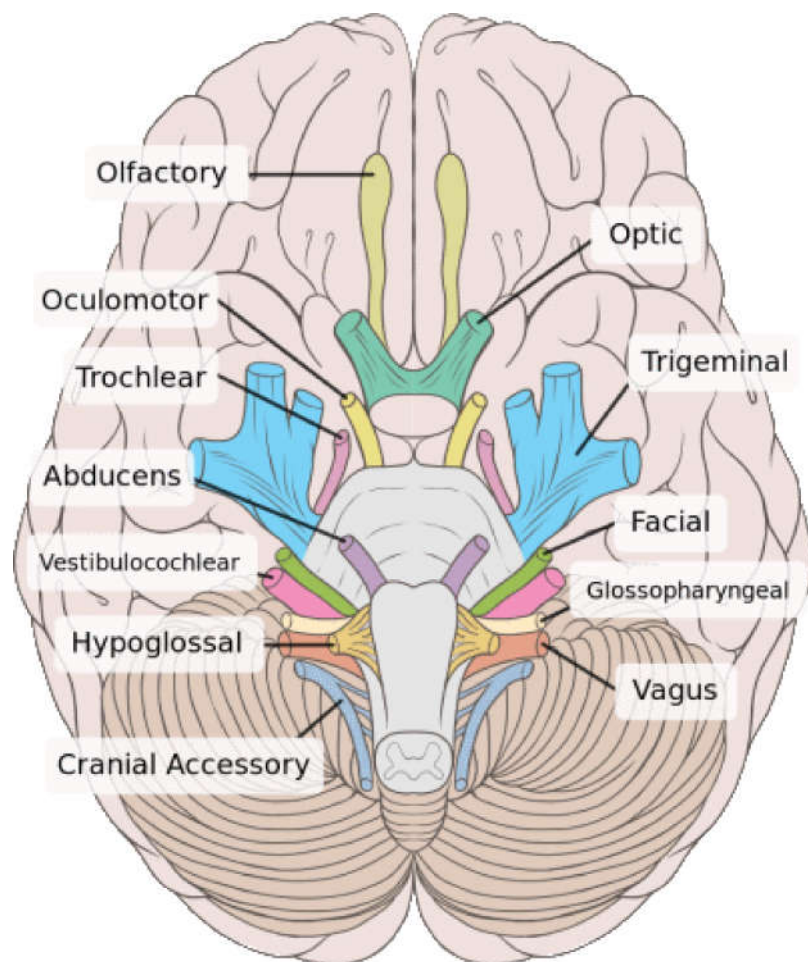


Image sourced from [Wikipedia](#)



View from the inferior surface of the brain showing the emergence of the cranial nerves

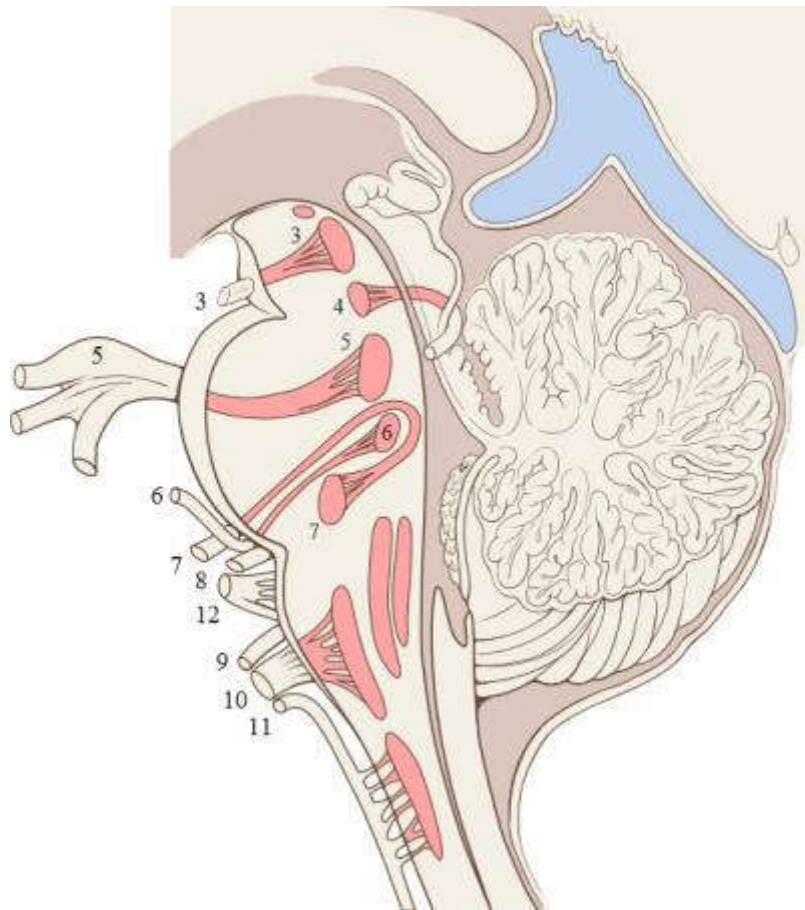


Image sourced from [Wikipedia](#)

Diagram showing the nuclei of the cranial nerves in the brainstem

Cranial nerve reflexes

Reflex	Afferent limb	Efferent limb
Corneal	Ophthalmic nerve (V ₁)	Facial nerve (VII)
Jaw jerk	Mandibular nerve (V ₃)	Mandibular nerve (V ₃)
Gag	Glossopharyngeal nerve (IX)	Vagal nerve (X)
Carotid sinus	Glossopharyngeal nerve (IX)	Vagal nerve (X)
Pupillary light	Optic nerve (II)	Oculomotor nerve (III)
Lacrimation	Ophthalmic nerve (V ₁)	Facial nerve (VII)

Creutzfeldt-Jakob disease

Creutzfeldt-Jakob disease (CJD) is rapidly progressive neurological condition caused by prion proteins. These proteins induce the formation of amyloid folds resulting in tightly packed beta-pleated sheets resistant to proteases.

Features

- dementia (rapid onset)
- myoclonus

Investigation

- CSF is usually normal
- EEG: biphasic, high amplitude sharp waves (only in sporadic CJD)
- MRI: hyperintense signals in the basal ganglia and thalamus

Sporadic CJD

- accounts for 85% of cases
- 10-15% of cases are familial
- mean age of onset is 65 years

New variant CJD

- younger patients (average age of onset = 25 years)
- psychological symptoms such as anxiety, withdrawal and dysphonia are the most common presenting features
- the 'prion protein' is encoded on chromosome 20 - it's role is not yet understood
- methionine homozygosity at codon 129 of the prion protein is a risk factor for developing CJD - all patients who have so far died have had this
- median survival = 13 months

Other prion diseases

- kuru
- fatal familial insomnia
- Gerstmann Straussler-Scheinker disease

Dementia

Dementia is thought to affect over 700,000 people in the UK and accounts for a large amount of health and social care spending. The most common cause of dementia in the UK is Alzheimer's disease followed by vascular and Lewy body dementia. These conditions may coexist.

Features

- diagnosis can be difficult and is often delayed
- assessment tools include the Abbreviated mental test score (AMTS), 6-Item cognitive impairment test (6CIT), General practitioner assessment of cognition (GPCOG) and the mini-mental state examination (MMSE) is widely used. A MMSE score of 24 or less out of 30 suggests dementia

Management

- in primary care a blood screen is usually sent to exclude reversible causes (e.g. Hypothyroidism). NICE recommend the following tests: FBC, U&E, LFTs, calcium, glucose, TFTs, vitamin B12 and folate levels. Patients are now commonly referred on to old-age psychiatrists (sometimes working in 'memory clinics').
- in secondary care neuroimaging is performed* to exclude other reversible conditions (e.g. Subdural haematoma, normal pressure hydrocephalus) and help provide information on aetiology to guide prognosis and management

*in the 2011 NICE guidelines structural imaging was said to be essential in the investigation of dementia

External Links:

[NICE](#)

2011 Dementia guidelines

[Alzheimers Society](#)

Helping you to assess cognition - A practical toolkit for clinicians

Dementia: causes

Common causes

- Alzheimer's disease
- cerebrovascular disease: multi-infarct dementia (c. 10-20%)
- Lewy body dementia (c. 10-20%)

Rarer causes (c. 5% of cases):

- Huntington's
- CJD
- Pick's disease (atrophy of frontal and temporal lobes)
- HIV (50% of AIDS patients)

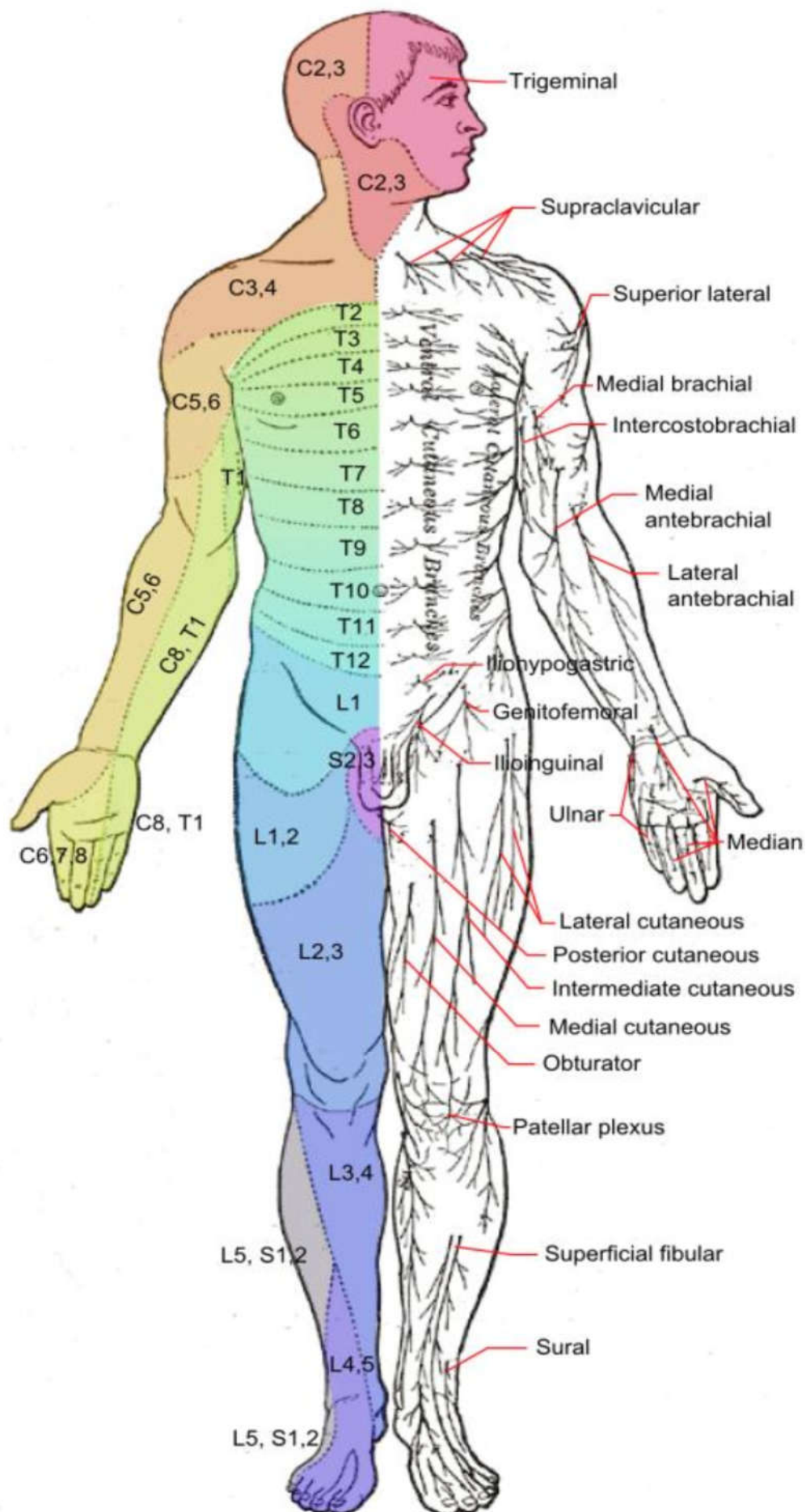
Important differentials, potentially treatable:

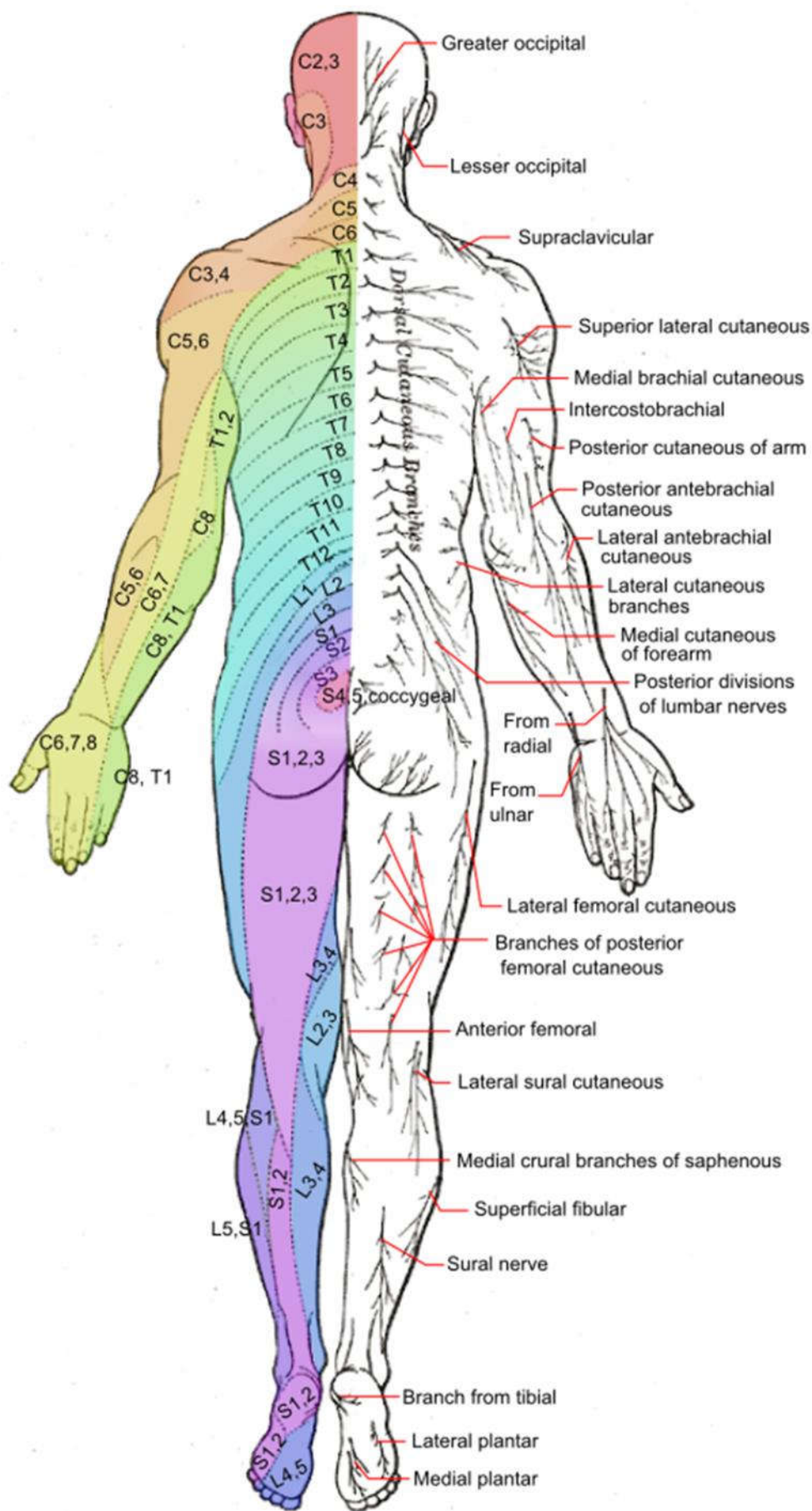
- hypothyroidism, Addison's
 - B12/folate/thiamine deficiency
 - syphilis
 - brain tumour
 - normal pressure hydrocephalus
 - subdural haematoma
 - depression
 - chronic drug use e.g. Alcohol, barbiturates
-

Dermatomes

The table below lists the major dermatome landmarks:

Nerve root	Landmark	Mnemonics
C2	Posterior half of the skull (cap)	
C3	High turtleneck shirt	
C4	Low-collar shirt	
C5, C6	Thumb + index finger	Make a 6 with your left hand by touching the tip of the thumb & index finger together - C6
C7	Middle finger + palm of hand	
C8	Ring + little finger	
T4	Nipples	T4 at the Teat Pore
T5	Inframammary fold	
T7	Xiphoid process	
T10	Umbilicus	BellybuT-TEN
L1	Inguinal ligament	L for ligament, 1 for Inguinal
L4	Knee caps	Down on aLL fours - L4
L5	Big toe, dorsum of foot (except lateral aspect)	L5 = Largest of the 5 toes
S1	Lateral foot, small toe	S1 = the smallest one
S2, S3	Genitalia	





Diabetes insipidus

Diabetes insipidus (DI) is a condition characterised by either a deficiency of antidiuretic hormone, ADH, (cranial DI) or an insensitivity to antidiuretic hormone (nephrogenic DI).

Causes of cranial DI

- idiopathic
- post head injury
- pituitary surgery
- craniopharyngiomas
- histiocytosis X
- DIDMOAD is the association of cranial Diabetes Insipidus, Diabetes Mellitus, Optic Atrophy and Deafness (also known as Wolfram's syndrome)

Causes of nephrogenic DI

- genetic: the more common form affects the vasopression (ADH) receptor, the less common form results from a mutation in the gene that encodes the aquaporin 2 channel
- electrolytes: hypercalcaemia, hypokalaemia
- drugs: demeclocycline, lithium
- tubulo-interstitial disease: obstruction, sickle-cell, pyelonephritis

Features

- polyuria
- polydipsia

Investigation

- high plasma osmolality, low urine osmolality
- water deprivation test

External Links:

[Merck manual](#)

Nephrogenic diabetes insipidus

Drugs causing peripheral neuropathy

Drugs causing a peripheral neuropathy:

- antibiotics: nitrofurantoin, metronidazole
- amiodarone
- isoniazid
- vincristine

DVLA: neurological disorders

The guidelines below relate to car/motorcycle use unless specifically stated. For obvious reasons, the rules relating to drivers of heavy goods vehicles tend to be much stricter

Specific rules

- first seizure: 6 months off driving*. For patients with established epilepsy they must be fit free for 12 months before being able to drive
- stroke or TIA: 1 month off driving, may not need to inform DVLA if no residual neurological deficit
- multiple TIAs over short period of times: 3 months off driving and inform DVLA
- craniotomy e.g. For meningioma: 1 year off driving**
- pituitary tumour: craniotomy: 6 months; trans-sphenoidal surgery 'can drive when there is no debarring residual impairment likely to affect safe driving'
- narcolepsy/cataplexy: cease driving on diagnosis, can restart once 'satisfactory control of symptoms'
- chronic neurological disorders e.g. multiple sclerosis, motor neuron disease: DVLA should be informed, complete PK1 form (application for driving licence holders state of health).

Syncope

- simple faint: no restriction
- single episode, explained and treated: 4 weeks off
- single episode, unexplained: 6 months off
- two or more episodes: 12 months off

*previously rule was 12 months. It is now 6 months off driving if the licence holder has undergone assessment by an appropriate specialist and no relevant abnormality has been identified on investigation, for example EEG and brain scan where indicated

**if the tumour is a benign meningioma and there is no seizure history, licence can be reconsidered 6 months after surgery if remains seizure free.

External Links:

[DVLA](#)

Neurological disorder guidelines

Eclampsia

Eclampsia may be defined as the development of seizures in association pre-eclampsia. **To recap, pre-eclampsia is defined as:**

- condition seen after 20 weeks gestation
- pregnancy-induced hypertension
- proteinuria

Magnesium sulphate is used to both prevent seizures in patients with severe pre-eclampsia and treat seizures once they develop. **Guidelines on its use suggest the following:**

- should be given once a decision to deliver has been made
- in eclampsia an IV bolus of 4g over 5-10 minutes should be given followed by an infusion of 1g / hour
- urine output, reflexes, respiratory rate and oxygen saturations should be monitored during treatment
- treatment should continue for 24 hours after last seizure or delivery (around 40% of seizures occur post-partum)

Other important aspects of treating severe pre-eclampsia/eclampsia include fluid restriction to avoid the potentially serious consequences of fluid overload

External Links:

[NICE](#)

2010 Hypertension in pregnancy: The management of hypertensive disorders during pregnancy

Epilepsy in children: syndromes

Infantile spasms (West's syndrome):

- brief spasms beginning in first few (4-6) months of life; M>F
 1. Flexion of head, trunk, limbs → extension of arms (Salaam attack); last 1-2 secs, repeat up to 50 times
 2. Progressive mental handicap
 3. EEG: hypsarrhythmia

Chapter: Neurology

- usually 2nd to serious neurological abnormality (e.g. TS, encephalitis, birth asphyxia) or may be cryptogenic
- poor prognosis
- vigabatrin/steroids

Typical (petit mal) absence seizures:

- onset 4-8 yrs
- duration few-30 secs; no warning, quick recovery; often many per day
- EEG: 3Hz generalized, symmetrical
- sodium valproate, ethosuximide
- good prognosis: 90-95% become seizure free in adolescence

Lennox-Gastaut syndrome:

- may be extension of infantile spasms (50% have hx)
- onset 1-5 yrs
- atypical absences, falls, jerks
- 90% moderate-severe mental handicap
- EEG: slow spike
- ketogenic diet may help

Benign rolandic epilepsy:

- most common in childhood, M>F
- paraesthesia (e.g. unilateral face), usually on waking up

Juvenile myoclonic epilepsy (Janz syndrome):

- onset: teens; F:M = 2:1
- 1. Infrequent generalized seizures, often in morning
- 2. Daytime absences
- 3. Sudden, shock like myoclonic seizure
- usually good response to sodium valproate

Neonatal period - try vitamin B6:

- 2nd: hypoglycaemia, meningitis, head trauma
- pyridoxine dependency (AR, IV B6)
- benign familial neonatal seizures (AD)
- benign neonatal convulsions (5th day)

Epilepsy: classification

Basics

- two main categories are generalised and partial seizures
- partial seizures may progress to general seizures
- other types: myoclonic, atypical absence, atonic and tonic seizures are usually seen in childhood

Generalised: - no focal features, consciousness lost immediately

- grand mal (tonic-clonic)
- petit mal (absence seizures)
- myoclonic: brief, rapid muscle jerks
- partial seizures progressing to generalised seizures.

Partial: - focal features depending on location

- simple (no disturbance of consciousness or awareness)
- complex (consciousness is disturbed)
- temporal lobe → aura, déjà vu, jamais vu; motor → Jacksonian

External Links:

[NICE](#)

2012 Epilepsy guidelines

Epilepsy: pregnancy and breast feeding

The risks of uncontrolled epilepsy during pregnancy generally outweigh the risks of medication to the fetus. All women thinking about becoming pregnant should be advised to take folic acid 5mg per day well before pregnancy to minimise the risk of neural tube defects. Around 1-2% of newborns born to non-epileptic mothers have congenital defects. This rises to 3-4% if the mother takes antiepileptic medication.

Other points:

- aim for monotherapy
- there is no indication to monitor antiepileptic drug levels
- sodium valproate: associated with neural tube defects
- carbamazepine: often considered the least teratogenic of the older antiepileptics
- phenytoin: associated with cleft palate
- lamotrigine: studies to date suggest the rate of congenital malformations may be low. The dose of lamotrigine may need to be increased in pregnancy

♦Breast feeding is generally considered safe for mothers taking antiepileptics with the possible exception of the barbiturates.

♦It is advised that pregnant women taking phenytoin are given vitamin K in the last month of pregnancy to prevent clotting disorders in the newborn.

Sodium valproate

The November 2013 issue of the Drug Safety Update also carried a warning about new evidence showing a significant risk of neurodevelopmental delay in children following maternal use of sodium valproate.

The update concludes that sodium valproate should not be used during pregnancy and in women of childbearing age unless clearly necessary. Women of childbearing age should not start treatment without specialist neurological or psychiatric advice.

External Links:

[NICE](#)

2012 Epilepsy guidelines

[SIGN](#)

2015 Diagnosis and management of epilepsy in adults

Epilepsy: treatment

Most neurologists now start antiepileptics following a second epileptic seizure. **NICE guidelines suggest starting antiepileptics after the first seizure if any of the following are present:**

- the patient has a neurological deficit
- brain imaging shows a structural abnormality
- the EEG shows unequivocal epileptic activity
- the patient or their family or carers consider the risk of having a further seizure unacceptable.

♦Sodium valproate is considered the first line treatment for patients with generalised seizures with carbamazepine used for partial seizures.

Generalized tonic-clonic seizures:

- sodium valproate
- second line: lamotrigine, carbamazepine

Absence seizures* (Petit mal):

- sodium valproate or ethosuximide
- sodium valproate particularly effective if co-existent tonic-clonic seizures in primary generalised epilepsy

Myoclonic seizures:

- sodium valproate
- second line: clonazepam, lamotrigine

Partial seizures

- carbamazepine or lamotrigine
- second line: sodium valproate

*carbamazepine may actually exacerbate absence seizure

External Links:

[NICE](#)

2012 Epilepsy guidelines

[Royal College of Physicians](#)

2013 Modern management of epilepsy.

Essential tremor

Essential tremor (previously called benign essential tremor) is an autosomal dominant condition which usually affects both upper limbs

Features

- postural tremor: worse if arms outstretched
- improved by alcohol and rest
- most common cause of titubation (head tremor)

Management

- propranolol is first-line
 - primidone is sometimes used
-

Ethosuximide

Ethosuximide is an antiepileptic that is particularly indicated in patients with absence seizures
Mechanism of action:

- blocks T-type calcium channels in thalamic neurons
-

Facial nerve

Supply - 'face, ear, taste, tear'

- face: muscles of facial expression
- ear: nerve to stapedius
- taste: supplies anterior two-thirds of tongue
- tear: parasympathetic fibres to lacrimal glands, also salivary glands

Causes of bilateral facial nerve palsy

- sarcoidosis
- Guillain-Barre syndrome
- polio, Lyme disease

Causes of unilateral facial nerve palsy - as above plus

Lower motor neuron	Upper motor neuron
• Bell's palsy • Ramsay-Hunt syndrome (due to herpes zoster) • acoustic neuroma • parotid tumours • HIV • multiple sclerosis* • diabetes mellitus	• stroke

LMN vs. UMN

- upper motor neuron lesion 'spares' upper face i.e. forehead
- lower motor neuron lesion affects all facial muscles

*may also cause an UMN palsy

Friedreich's ataxia

♦Friedreich's ataxia is the most common of the early-onset hereditary ataxias. It is an autosomal recessive, trinucleotide repeat disorder characterised by a GAA repeat in the X25 gene on chromosome 9 (frataxin). Friedreich's ataxia is unusual amongst trinucleotide repeat disorders in not demonstrating the phenomenon of anticipation.

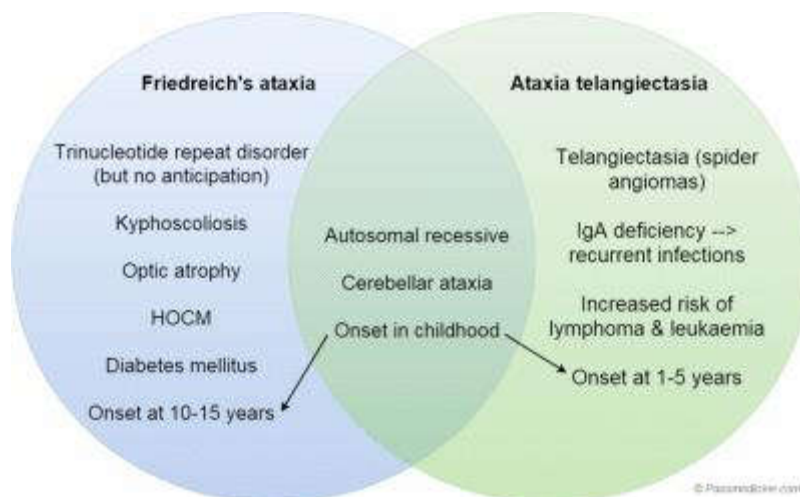
♦The typical age of onset is 10-15 years old. Gait ataxia and kyphoscoliosis are the most common presenting features.

Neurological features:

- absent ankle jerks/extensor plantars
- cerebellar ataxia
- optic atrophy
- spinocerebellar tract degeneration

Other features

- hypertrophic obstructive cardiomyopathy (90%, most common cause of death)
- diabetes mellitus (10-20%)
- high-arched palate.



Comparison of Friedrich's ataxia and ataxic telangiectasia. Note in particular how ataxic telangiectasia tends to present much earlier, often at the age of 1-2 years

Frontotemporal lobar degeneration

Frontotemporal lobar degeneration (FTLD) is the third most common type of cortical dementia after Alzheimer's and Lewy body dementia.

There are three recognised types of FTLD:

- Frontotemporal dementia (Pick's disease).
- Progressive non fluent aphasia (chronic progressive aphasia, CPA).
- Semantic dementia.

Common features of frontotemporal lobar dementias

Onset before 65
Insidious onset
Relatively preserved memory and visuospatial skills
Personality change and social conduct problems

Pick's disease

◆ This is the most common type and is characterised by personality change and impaired social conduct. Other common features include hyperorality, disinhibition, increased appetite, and perseveration behaviours.

◆ Focal gyral atrophy with a knife-blade appearance is characteristic of Pick's disease.

◆ Macroscopic changes seen in Pick's disease include:-

- Atrophy of the frontal and temporal lobes

♦**Microscopic changes include:-**

- Pick bodies - spherical aggregations of tau protein (silver-staining)
- Gliosis
- Neurofibrillary tangles
- Senile plaques

CPA

Here the chief factor is non fluent speech. They make short utterances that are agrammatic. Comprehension is relatively preserved.

Semantic dementia

Here the patient has a fluent progressive aphasia. The speech is fluent but empty and conveys little meaning. Unlike in Alzheimer's memory is better for recent rather than remote events.

Guillain-Barre syndrome

Guillain-Barre syndrome describes an immune mediated demyelination of the peripheral nervous system often triggered by an infection (classically *Campylobacter jejuni*)

Pathogenesis:

- cross reaction of antibodies with gangliosides in the peripheral nervous system
- correlation between anti-ganglioside antibody (e.g. anti-GM1) and clinical features has been demonstrated
- anti-GM1 antibodies in 25% of patients

Miller Fisher syndrome

- variant of Guillain-Barre syndrome
 - associated with ophthalmoplegia, areflexia and ataxia. The eye muscles are typically affected first
 - usually presents as a descending paralysis rather than ascending as seen in other forms of Guillain-Barre syndrome
 - anti-GQ1b antibodies are present in 90% of cases
-

Guillain-Barre syndrome: features

Guillain-Barre syndrome describes an immune mediated demyelination of the peripheral nervous system often triggered by an infection (classically *Campylobacter jejuni*).

The characteristic features of Guillain-Barre syndrome is progressive weakness of all four limbs. The weakness is classically ascending i.e. the lower extremities are affected first, however it tends to affect proximal muscles earlier than the distal ones. Sensory symptoms tend to be mild (e.g. distal paraesthesia) with very few sensory signs. Some patients experience back pain in the initial stages of the illness

Other features:

- areflexia
- cranial nerve involvement e.g. diplopia
- autonomic involvement: e.g. urinary retention

Less common findings

- papilloedema: thought to be secondary to reduced CSF resorption

External Links:

[Patient.info](#)

Guillain-Barre syndrome review

Guillain-Barre syndrome: management

Guillain-Barre syndrome describes an immune mediated demyelination of the peripheral nervous system often triggered by an infection (classically *Campylobacter jejuni*).

Management:

- plasma exchange
- IV immunoglobulins (IVIG): as effective as plasma exchange. No benefit in combining both treatments. IVIG may be easier to administer and tends to have fewer side-effects
- steroids and immunosuppressants have not been shown to be beneficial
- FVC regularly to monitor respiratory function

Prognosis

- 20% suffer permanent disability, 5% die

Guillain-Barre syndrome: prognosis

Guillain-Barre syndrome (GBS) describes an immune mediated demyelination of the peripheral nervous system often triggered by an infection (classically *Campylobacter jejuni*)

Poor prognostic features:

- age > 40 years
 - poor upper extremity muscle strength
 - previous history of a diarrhoeal illness (specifically *Campylobacter jejuni*)
 - high anti-GM1 antibody titre
 - need for ventilatory support
- There is currently contradictory evidence as to whether a gradual or rapid onset of GBS is associated with a poor outcome

Head injury: NICE guidance

NICE has strict and clear guidance regarding which adult patients are safe to discharge and which need further CT head imaging. **The latter group are also divided into two further cohorts, those who require an immediate CT head and those requiring CT head within 8 hours of injury:**

CT head immediately

- GCS < 13 on initial assessment
- GCS < 15 at 2 hours post-injury
- suspected open or depressed skull fracture.
- any sign of basal skull fracture (haemotympanum, 'panda' eyes, cerebrospinal fluid leakage from the ear or nose, Battle's sign).
- post-traumatic seizure.
- focal neurological deficit.
- more than 1 episode of vomiting.

CT head scan within 8 hours of the head injury - for adults with any of the following risk factors who have experienced some loss of consciousness or amnesia since the injury:

- age 65 years or older
- any history of bleeding or clotting disorders
- dangerous mechanism of injury (a pedestrian or cyclist struck by a motor vehicle, an occupant ejected from a motor vehicle or a fall from a height of greater than 1 metre or 5 stairs)
- more than 30 minutes' retrograde amnesia of events immediately before the head injury

♦If a patient is on warfarin who have sustained a head injury with no other indications for a CT head scan, perform a CT head scan within 8 hours of the injury.

External Links:

[NICE](#)

2014 Head injury guidelines

Head injury: types of traumatic brain injury

Basics

- primary brain injury may be focal (contusion/haematoma) or diffuse (diffuse axonal injury)
- diffuse axonal injury occurs as a result of mechanical shearing following deceleration, causing disruption and tearing of axons
- intra-cranial haematomas can be extradural, subdural or intracerebral, while contusions may occur adjacent to (coup) or contralateral (contre-coup) to the side of impact
- secondary brain injury occurs when cerebral oedema, ischaemia, infection, tonsillar or tentorial herniation exacerbates the original injury. The normal cerebral autoregulatory processes are disrupted following trauma rendering the brain more susceptible to blood flow changes and hypoxia
- the Cushing's reflex (hypertension and bradycardia) often occurs late and is usually a pre terminal event.

Type of injury	Notes
Extradural (epidural) haematoma	Bleeding into the space between the dura mater and the skull. Often results from acceleration-deceleration trauma or a blow to the side of the head. The majority of extradural haematomas occur in the temporal region where skull fractures cause a rupture of the middle meningeal artery. Features • features of raised intracranial pressure • some patients may exhibit a lucid interval
Subdural haematoma	Bleeding into the outermost meningeal layer. Most commonly occur around the frontal and parietal lobes. Risk factors include old age, alcoholism and anticoagulation. Slower onset of symptoms than a extradural haematoma.
Subarachnoid haemorrhage	Usually occurs spontaneously in the context of a ruptured cerebral aneurysm but may be seen in association with other injuries when a patient has sustained a traumatic brain injury

Image gallery

Extradural (epidural) haematoma:

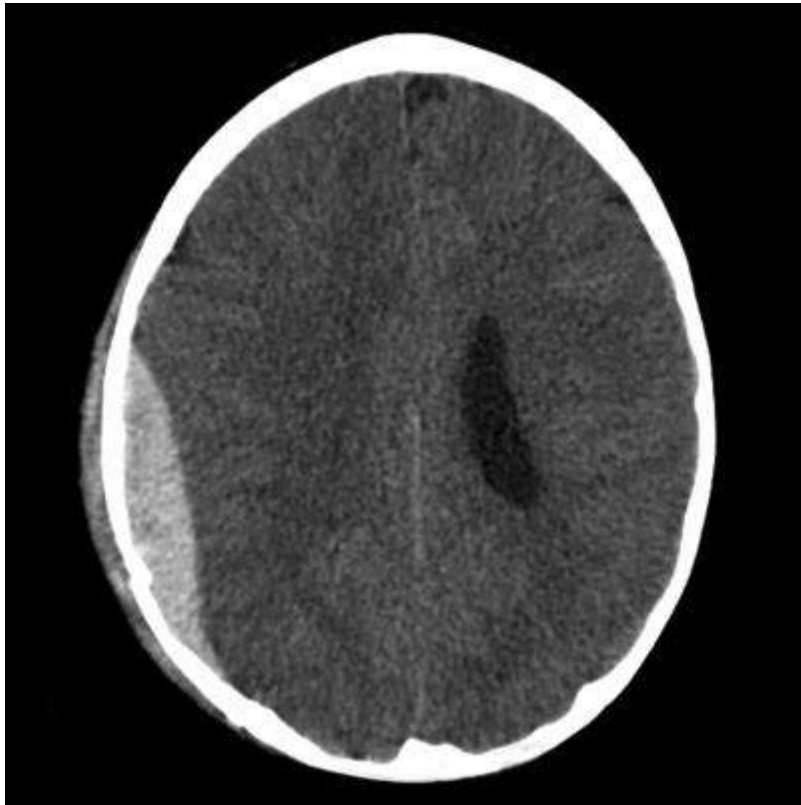


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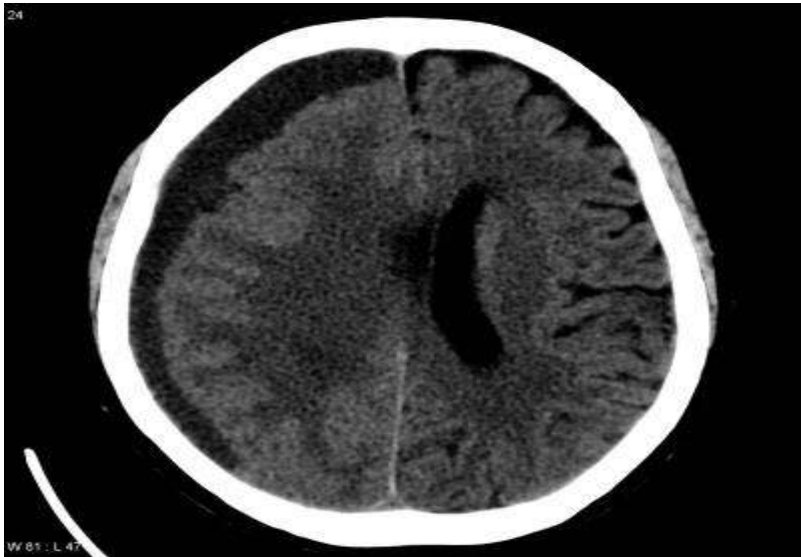




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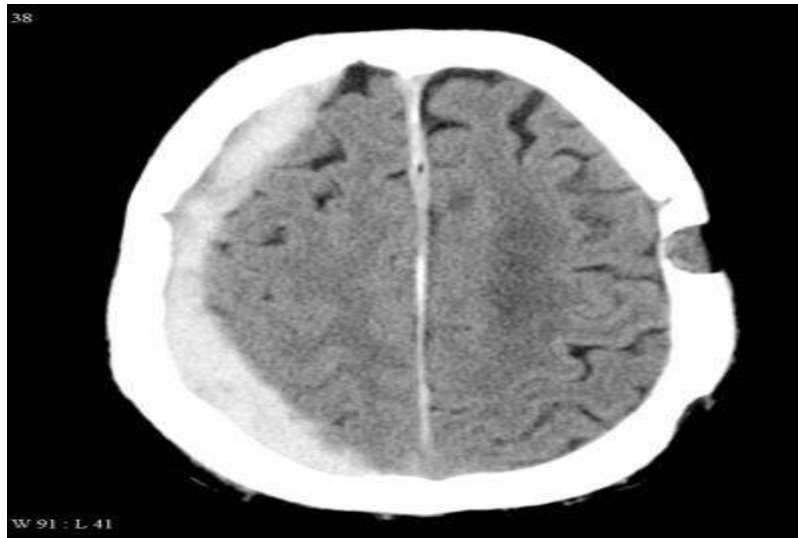


Subdural haematoma:



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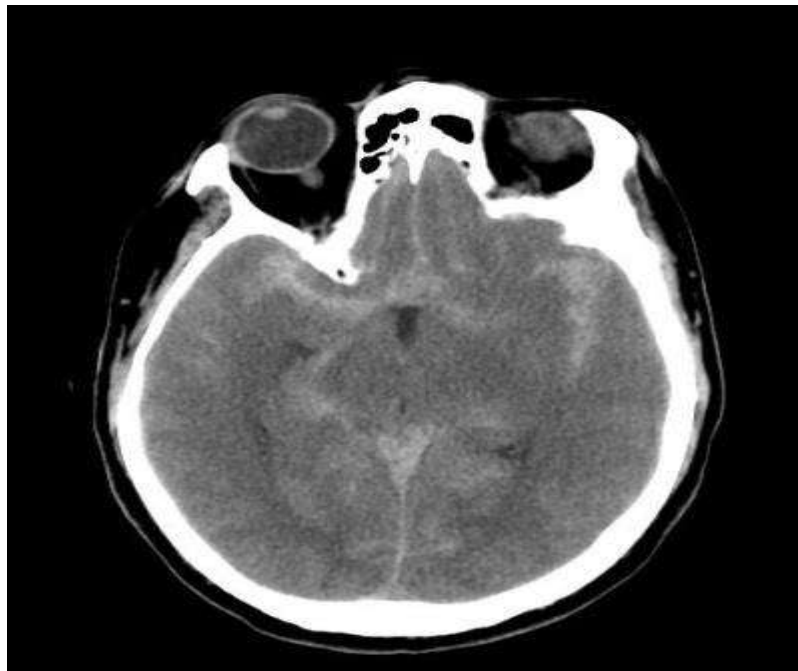




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Subarachnoid haemorrhage:



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External Links:

[NICE](#)

2014 Head injury guidelines

Headache

Headache accounts for a large proportion of medical consultations. **The table below summarises the main characteristics of common or important causes:**

Migraine	Recurrent, severe headache which is usually unilateral and throbbing in nature May be associated with aura, nausea and photosensitivity Aggravated by, or causes avoidance of, routine activities of daily living. Patients often describe 'going to bed'. In women may be associated with menstruation
Tension headache	Recurrent, non-disabling, bilateral headache, often described as a 'tight-band' Not aggravated by routine activities of daily living
Cluster headache*	Pain typical occurs once or twice a day, each episode lasting 15 mins - 2 hours with clusters typically lasting 4-12 weeks Intense pain around one eye (recurrent attacks 'always' affect same side) Patient is restless during an attack Accompanied by redness, lacrimation, lid swelling More common in men and smokers
Temporal arteritis	Typically patient > 60 years old Usually rapid onset (e.g. < 1 month) of unilateral headache Jaw claudication (65%) Tender, palpable temporal artery Raised ESR
Medication overuse headache	Present for 15 days or more per month Developed or worsened whilst taking regular symptomatic medication Patients using opioids and triptans are at most risk May be psychiatric co-morbidity

Other causes of headache:

Acute single episode:

- meningitis
- encephalitis
- subarachnoid haemorrhage
- head injury
- sinusitis

- glaucoma (acute closed-angle)
- tropical illness e.g. Malaria

Chronic headache

- chronically raised ICP
- Paget's disease
- psychological

*some neurologists use the term trigeminal autonomic cephalgia to group a number of conditions including cluster headache, paroxysmal hemicrania and short-lived unilateral neuralgiform headache with conjunctival injection and tearing (SUNCT). It is recommended such patients are referred for specialist assessment as specific treatment may be required, for example it is known paroxysmal hemicrania responds very well to indomethacin

External Links:

[NICE](#)

2012 Headaches: Diagnosis and management of headaches in young people and adults

[SIGN](#)

2008 Diagnosis and management of headache in adults

Hemiballism

♦Hemiballism occurs following damage to the subthalamic nucleus. Ballistic movements are involuntary, sudden, jerking movements which occur contralateral to the side of the lesion. The ballistic movements primarily affect the proximal limb musculature whilst the distal muscles may display more choreiform-like movements

♦Symptoms may decrease whilst the patient is asleep.

♦Antidopaminergic agents (e.g. Haloperidol) are the mainstay of treatment

Herpes simplex encephalitis

Herpes simplex (HSV) encephalitis is a common topic in the exam. The virus characteristically affects the temporal lobes - questions may give the result of imaging or describe temporal lobe signs e.g. aphasia

Features

- fever, headache, psychiatric symptoms, seizures, vomiting
- focal features e.g. aphasia
- peripheral lesions (e.g. cold sores) have no relation to presence of HSV encephalitis

Pathophysiology

- HSV-1 responsible for 95% of cases in adults
- typically affects temporal and inferior frontal lobes

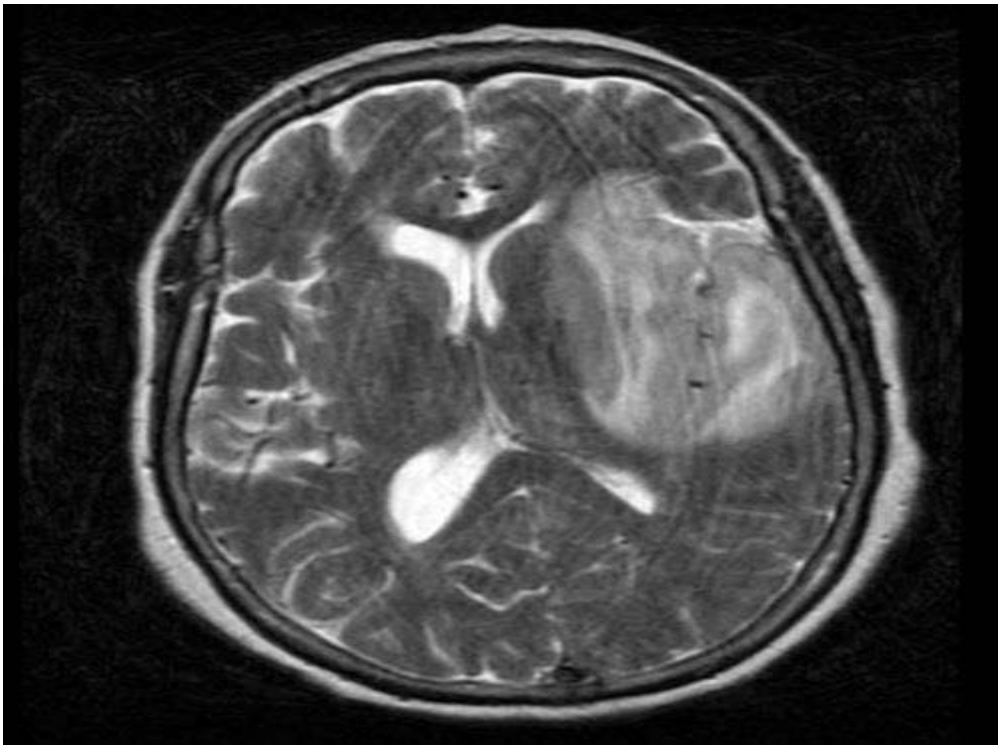
Investigation

- CSF: lymphocytosis, elevated protein
- PCR for HSV
- CT: medial temporal and inferior frontal changes (e.g. petechial haemorrhages) - normal in one-third of patients
- MRI is better
- EEG pattern: lateralised periodic discharges at 2 Hz

Treatment

- intravenous aciclovir

♦The prognosis is dependent on whether aciclovir is commenced early. If treatment is started promptly the mortality is 10-20%. Left untreated the mortality approaches 80%.



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MRI of a patient with HSV encephalitis. There is hyperintensity of the affected white matter and cortex in the medial temporal lobes and insular cortex.

Hodgkin's lymphoma: histological classification and prognosis

Hodgkin's lymphoma is a malignant proliferation of lymphocytes characterised by the presence of the Reed-Sternberg cell. It has a bimodal age distributions being most common in the third and seventh decades

Histological classification

Type	Frequency	Prognosis	Notes
Nodular sclerosing	Most common (around 70%)	Good prognosis	More common in women. Associated with lacunar cells
Mixed cellularity	Around 20%	Good prognosis	Associated with a large number of Reed-Sternberg cells
Lymphocyte predominant	A*round 5%	Best prognosis	
Lymphocyte depleted	Rare	Worst prognosis	

'B' symptoms also imply a poor prognosis:

- weight loss > 10% in last 6 months
- fever > 38°C
- night sweats

Other factors associated with a poor prognosis identified in a 1998 NEJM paper included:

- age > 45 years
- stage IV disease
- haemoglobin < 10.5 g/dl
- lymphocyte count < 600/ μ l or < 8%
- male
- albumin < 40 g/l
- white blood count > 15,000/ μ l

*Reed-Sternberg cells with nuclei surrounded by a clear space

Horner's syndrome

Features:

- miosis (small pupil)
- ptosis
- enophthalmos* (sunken eye)
- anhidrosis (loss of sweating one side)

Distinguishing between causes:

- heterochromia (difference in iris colour) is seen in congenital Horner's
- anhidrosis: see below

Central lesions	Pre-ganglionic lesions	Post-ganglionic lesions
Anhidrosis of the face, arm and trunk	Anhidrosis of the face	No anhidrosis
Stroke Syringomyelia Multiple sclerosis Tumour Encephalitis	Pancoast's tumour Thyroidectomy Trauma Cervical rib	Carotid artery dissection Carotid aneurysm Cavernous sinus thrombosis Cluster headache

*in reality the appearance is due to a narrow palpebral aperture rather than true enophthalmos

HSMN

Hereditary sensorimotor neuropathy (HSMN) is a relatively new term which encompasses Charcot-Marie-Tooth disease (also known as peroneal muscular atrophy). Over 7 types have been characterised - however only 2 are common to clinical practice

- HSMN type I: primarily due to demyelinating pathology
- HSMN type II: primarily due to axonal pathology

HSMN type I

- autosomal dominant
- due to defect in PMP-22 gene (which codes for myelin)
- features often start at puberty
- motor symptoms predominate
- distal muscle wasting, pes cavus, clawed toes
- foot drop, leg weakness often first features

Huntington's disease

Huntington's disease is an inherited neurodegenerative condition. It is a progressive and incurable condition that typically results in death 20 years after the initial symptoms develop.

Genetics

- autosomal dominant
- trinucleotide repeat disorder: repeat expansion of CAG
- results in degeneration of cholinergic and GABAergic neurons in the striatum of the basal ganglia
- due to defect in huntingtin gene on chromosome 4

Features typical develop after 35 years of age

- chorea
- personality changes (e.g. irritability, apathy, depression) and intellectual impairment
- dystonia
- saccadic eye movements

Idiopathic intracranial hypertension

Idiopathic intracranial hypertension (also known as pseudotumour cerebri and formerly benign intracranial hypertension) is a condition classically seen in young, overweight females.

Features

- headache
- blurred vision
- papilloedema (usually present)
- enlarged blind spot
- sixth nerve palsy may be present

Risk factors

- obesity
- female sex
- pregnancy
- drugs*: oral contraceptive pill, steroids, tetracycline, vitamin A, lithium

Management

- weight loss
- diuretics e.g. acetazolamide
- topiramate is also used, and has the added benefit of causing weight loss in most patients
- repeated lumbar puncture
- surgery: optic nerve sheath decompression and fenestration may be needed to prevent damage to the optic nerve. A lumboperitoneal or ventriculoperitoneal shunt may also be performed to reduce intracranial pressure

*if intracranial hypertension is thought to occur secondary to a known causes (e.g. Medication) then it is of course not idiopathic

External Links:

[Patient.info](#)

Idiopathic intracranial hypertension review.

Intracranial venous thrombosis

Overview

- can cause cerebral infarction, much less common than arterial causes
- 50% of patients have isolated sagittal sinus thromboses - the remainder have coexistent lateral sinus thromboses and cavernous sinus thromboses

Features

- headache (may be sudden onset)
- nausea & vomiting.

Sagittal sinus thrombosis

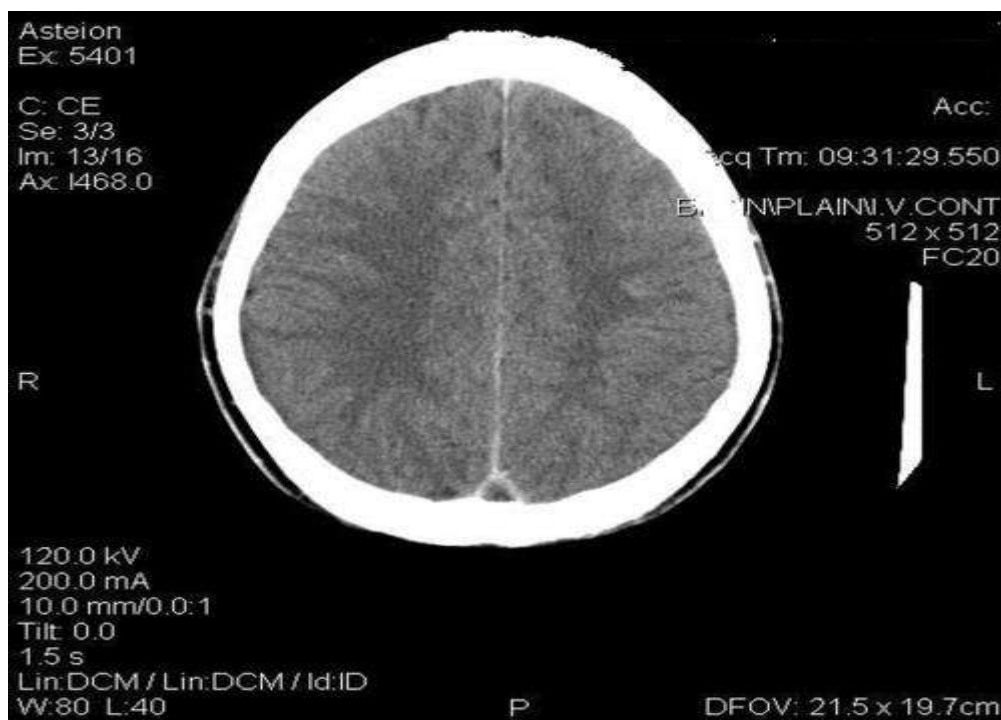
- may present with seizures and hemiplegia
- parasagittal biparietal or bifrontal haemorrhagic infarctions are sometimes seen.

Cavernous sinus thrombosis

- other causes of cavernous sinus syndrome: local infection (e.g. sinusitis), neoplasia, trauma
- periorbital oedema
- ophthalmoplegia: 6th nerve damage typically occurs before 3rd & 4th
- trigeminal nerve involvement may lead to hyperaesthesia of upper face and eye pain
- central retinal vein thrombosis

Lateral sinus thrombosis

- 6th and 7th cranial nerve palsies



© Image used on license from [Radiopaedia](#)

CT with contrast demonstrating a **superior sagittal sinus thrombosis** showing the typical empty delta sign. Look at the 'bottom' of the scan for the triangular shaped dural sinus. This should normally be white due to it being filled with contrast. The empty delta sign occurs when the thrombus fails to enhance within the dural sinus and is outlined by enhanced collateral channels in the falx. This sign is seen in only about 25%-30% of cases but is highly diagnostic for sagittal sinus thrombosis

External Links:

[Webmed Central](#)

Diagram of types of intracranial venous thrombosis

Lambert-Eaton syndrome

Lambert-Eaton myasthenic syndrome is seen in association with small cell lung cancer, and to a lesser extent breast and ovarian cancer. It may also occur independently as an autoimmune disorder. Lambert-Eaton myasthenic syndrome is caused by an antibody directed against pre-synaptic voltage gated calcium channel in the peripheral nervous system

Features:

- repeated muscle contractions lead to increased muscle strength* (in contrast to myasthenia gravis)
- limb girdle weakness (affects lower limbs first)
- hyporeflexia
- autonomic symptoms: dry mouth, impotence, difficulty micturating
- ophthalmoplegia and ptosis not commonly a feature (unlike in myasthenia gravis)

EMG

- incremental response to repetitive electrical stimulation

Management

- treatment of underlying cancer
- immunosuppression, for example with prednisolone and/or azathioprine
- 3,4-diaminopyridine is currently being trialled**
- intravenous immunoglobulin therapy and plasma exchange may be beneficial

*in reality this is seen in only 50% of patients and following prolonged muscle use muscle strength will eventually decrease

**works by blocking potassium channel efflux in the nerve terminal so that the action potential duration is increased. Calcium channels can then be open for a longer time and allow greater acetylcholine release to the stimulate muscle at the end plate

Lateral medullary syndrome

Lateral medullary syndrome, also known as Wallenberg's syndrome, occurs following occlusion of the posterior inferior cerebellar artery

Cerebellar features

- ataxia
- nystagmus

Brainstem features

- ipsilateral: dysphagia, facial numbness, cranial nerve palsy e.g. Horner's
 - contralateral: limb sensory loss
-

Levodopa

Overview

- usually combined with a decarboxylase inhibitor (e.g. carbidopa or benserazide) to prevent peripheral metabolism of L-dopa to dopamine
- reduced effectiveness with time (usually by 2 years)
- no use in neuroleptic induced parkinsonism

Adverse effects

- dyskinesia
 - 'on-off' effect
 - postural hypotension
 - cardiac arrhythmias
 - nausea & vomiting
 - psychosis
 - reddish discolouration of urine upon standing
-

Lewy body dementia

♦Lewy body dementia is an increasingly recognised cause of dementia, accounting for up to 20% of cases. The characteristic pathological feature is alpha-synuclein cytoplasmic inclusions (Lewy bodies) in the substantia nigra, paralimbic and neocortical areas

♦The relationship between Parkinson's disease and Lewy body dementia is complicated, particularly as dementia is often seen in Parkinson's disease. Also, up to 40% of patients with Alzheimer's have Lewy bodies

♦Neuroleptics should be avoided in Lewy body dementia as patients are extremely sensitive and may develop irreversible parkinsonism. Questions may give a history of a patient who has deteriorated following the introduction of an antipsychotic agent

Features:

- progressive cognitive impairment
- parkinsonism
- visual hallucinations (other features such as delusions and non-visual hallucinations may also be seen)

Diagnosis

- usually clinical
- single-photon emission computed tomography (SPECT) is increasingly used. It is currently commercially known as a DaTscan. Dopaminergic iodine-123-radiolabelled 2-carbomethoxy-3-(4-iodophenyl)-N-(3-fluoropropyl) nortropane (123-I FP-CIT) is used as the radioisotope. The sensitivity of SPECT in diagnosing Lewy body dementia is around 90% with a specificity of 100%

Medication overuse headache

Medication overuse headache is one of the most common causes of chronic daily headache. It may affect up to 1 in 50 people

Features:

- present for 15 days or more per month
- developed or worsened whilst taking regular symptomatic medication
- patients using opioids and triptans are at most risk
- may be psychiatric co-morbidity

Management (from 2008 SIGN guidelines):

- simple analgesics and triptans should be withdrawn abruptly (may initially worsen headaches)
- opioid analgesics should be gradually withdrawn

External Links:

[SIGN](#)

2008 Diagnosis and management of headache in adults.

Meniere's disease

Meniere's disease is a disorder of the inner ear of unknown cause. It is characterised by excessive pressure and progressive dilation of the endolymphatic system. It is more common in middle-aged adults but may be seen at any age. Meniere's disease has a similar prevalence in both men and women.

Features:

- recurrent episodes of vertigo, tinnitus and hearing loss (sensorineural). Vertigo is usually the prominent symptom
- a sensation of aural fullness or pressure is now recognised as being common
- other features include nystagmus and a positive Romberg test
- episodes last minutes to hours
- typically symptoms are unilateral but bilateral symptoms may develop after a number of years.

Natural history:

- symptoms resolve in the majority of patients after 5-10 years
- the majority of patients will be left with a degree of hearing loss
- psychological distress is common

Management

- ENT assessment is required to confirm the diagnosis
- patients should inform the DVLA. The current advice is to cease driving until satisfactory control of symptoms is achieved
- acute attacks: buccal or intramuscular prochlorperazine. Admission is sometimes required
- prevention: betahistine may be of benefit

External Links:

[Clinical Knowledge Summaries](#)

Meniere's disease guidelines.

Meralgia paraesthetica

Basics:

- caused by compression of lateral cutaneous nerve of thigh
 - typically burning sensation over antero-lateral aspect of thigh
-

Migraine: diagnostic criteria

The International Headache Society has produced the following diagnostic criteria for migraine without aura:

Point	Criteria
A	At least 5 attacks fulfilling criteria B-D
B	Headache attacks lasting 4-72 hours* (untreated or unsuccessfully treated)
C	Headache has at least two of the following characteristics: 1. unilateral location* 2. pulsating quality (i.e., varying with the heartbeat) 3. moderate or severe pain intensity 4. aggravation by or causing avoidance of routine physical activity (e.g., walking or climbing stairs)
D	During headache at least one of the following: 1. nausea and/or vomiting* 2. photophobia and phonophobia
E	Not attributed to another disorder (history and examination do not suggest a secondary headache disorder or, if they do, it is ruled out by appropriate investigations or headache attacks do not occur for the first time in close temporal relation to the other disorder)

*In children, attacks may be shorter-lasting, headache is more commonly bilateral, and gastrointestinal disturbance is more prominent.

Migraine with aura (seen in around 25% of migraine patients) tends to be easier to diagnose with a typical aura being progressive in nature and may occur hours prior to the headache. Typical aura include a transient hemianopic disturbance or a spreading scintillating scotoma ('jagged crescent'). Sensory symptoms may also occur.

♦If we compare these guidelines to the NICE criteria the following points are noted:

- NICE suggests migraines may be unilateral or bilateral
- NICE also give more detail about typical auras:

♦Auras may occur with or without headache and:

- are fully reversible.
- develop over at least 5 minutes.
- last 5-60 minutes.

♦**The following aura symptoms are atypical and may prompt further investigation/referral:**

- motor weakness
- double vision
- visual symptoms affecting only one eye
- poor balance
- decreased level of consciousness.

External Links:

[British Association for the Study of Headache](#)

Headache guidelines

Migraine: pregnancy, contraception and other hormonal factors

SIGN produced guidelines in 2008 on the management of migraine, the following is selected highlights:

Migraine during pregnancy

- paracetamol 1g is first-line
- aspirin 300mg or ibuprofen 400mg can be used second-line in the first and second trimester

Migraine and the combined oral contraceptive (COC) pill:

- if patients have migraine with aura then the COC is absolutely contraindicated due to an increased risk of stroke (relative risk 8.72)

Migraine and menstruation

- many women find that the frequency and severity of migraines increase around the time of menstruation
- SIGN recommends that women are treated with mefenamic acid or a combination of aspirin, paracetamol and caffeine. Triptans are also recommended in the acute situation

Migraine and hormone replacement therapy (HRT)

- safe to prescribe HRT for patients with a history of migraine but it may make migraines worse

External Links:

[SIGN](#)

2008 Diagnosis and management of headache in adults

Miosis

Causes of miosis (small pupil):

- Horner's syndrome
- Argyll-Robertson pupil
- senile miosis
- pontine haemorrhage
- congenital

Drugs causes

- opiates
- parasympathomimetics: pilocarpine
- organophosphate toxicity

Motor neuron disease: features

Motor neuron disease is a neurological condition of unknown cause which can present with both upper and lower motor neuron signs. It rarely presents before 40 years and various patterns of disease are recognised including amyotrophic lateral sclerosis, progressive muscular atrophy and bulbar palsy

There are a number of clues which point towards a diagnosis of motor neuron disease:

- fasciculation
- the absence of sensory signs/symptoms*
- the mixture of lower motor neurone and upper motor neurone signs
- wasting of the small hand muscles/tibialis anterior is common

Other features:

- doesn't affect external ocular muscles
- no cerebellar signs
- abdominal reflexes are usually preserved and sphincter dysfunction if present is a late feature

♦The diagnosis of motor neuron disease is **clinical**, but nerve conduction studies will show normal motor conduction and can help exclude a neuropathy. Electromyography shows a reduced number of action potentials with an increased amplitude. MRI is usually performed to exclude the differential diagnosis of cervical cord compression and myelopathy

*vague sensory symptoms may occur early in the disease (e.g. limb pain) but 'never' sensory signs

External Links:

[Royal College of Physicians](#)

Motor neuron disease: diagnostic pitfalls

[Postgraduate Medical Journal](#)

Review of MND

Motor neuron disease: management

Motor neuron disease is a neurological condition of unknown cause which can present with both upper and lower motor neuron signs. It rarely presents before 40 years and various patterns of disease are recognised including amyotrophic lateral sclerosis, progressive muscular atrophy and bulbar palsy

Riluzole

- prevents stimulation of glutamate receptors
- used mainly in amyotrophic lateral sclerosis
- prolongs life by about 3 months

Respiratory care

- non-invasive ventilation (usually BIPAP) is used at night
- studies have shown a survival benefit of around 7 months

Prognosis

- poor: 50% of patients die within 3 years

External Links:

[NICE](#)

2016 Motor neurone disease: assessment and management

Motor neuron disease: types

Motor neuron disease is a neurological condition of unknown cause which can present with both upper and lower motor neuron signs. It rarely presents before 40 years and various patterns of disease are recognised including amyotrophic lateral sclerosis, primary lateral sclerosis, progressive muscular atrophy and progressive bulbar palsy. In some patients however, there is a combination of clinical patterns

Amyotrophic lateral sclerosis (50% of patients):

- typically LMN signs in arms and UMN signs in legs
- in familial cases the gene responsible lies on chromosome 21 and codes for superoxide dismutase

Primary lateral sclerosis

- UMN signs only

Progressive muscular atrophy

- LMN signs only
- affects distal muscles before proximal
- carries best prognosis

Progressive bulbar palsy

- palsy of the tongue, muscles of chewing/swallowing and facial muscles due to loss of function of brainstem motor nuclei
- carries worst prognosis

External Links:

[Postgraduate Medical Journal](#)

Review of MND

Multiple sclerosis: features

Patient's with multiple sclerosis (MS) may present with non-specific features, for example around 75% of patients have significant lethargy.

Visual

- optic neuritis: common presenting feature
- optic atrophy
- Uhthoff's phenomenon: worsening of vision following rise in body temperature
- internuclear ophthalmoplegia

Sensory

- pins/needles
- numbness
- trigeminal neuralgia
- Lhermitte's syndrome: paraesthesiae in limbs on neck flexion

Motor

- spastic weakness: most commonly seen in the legs

Cerebellar

- ataxia: more often seen during an acute relapse than as a presenting symptom
- tremor

Others

- urinary incontinence
 - sexual dysfunction
 - intellectual deterioration
-

External Links:

[NICE](#)

2014 Multiple sclerosis guidelines

Multiple sclerosis: management

Treatment in multiple sclerosis is focused at reducing the frequency and duration of relapses. There is no cure.

Acute relapse

High dose steroids (e.g. oral or IV methylprednisolone) may be given for 5 days to shorten the length of an acute relapse. It should be noted that steroids shorten the duration of a relapse and do not alter the degree of recovery (i.e. whether a patient returns to baseline function)

Disease modifying drugs

Beta-interferon has been shown to reduce the relapse rate by up to 30%. Certain criteria have to be met before it is used:

- relapsing-remitting disease + 2 relapses in past 2 years + able to walk 100m unaided
- secondary progressive disease + 2 relapses in past 2 years + able to walk 10m (aided or unaided)
- reduces number of relapses and MRI changes, however doesn't reduce overall disability

Other drugs used in the management of multiple sclerosis include:

- glatiramer acetate: immunomodulating drug - acts as an 'immune decoy'
- natalizumab: a recombinant monoclonal antibody that antagonises Alpha-4 Beta-1-integrin found on the surface of leucocytes, thus inhibiting migration of leucocytes across the endothelium across the blood-brain barrier
- fingolimod: sphingosine 1-phosphate receptor modulator, prevents lymphocytes from leaving lymph nodes. An oral formulation is available

Some specific problems

Fatigue

- once other problems (e.g. anaemia, thyroid or depression) have been excluded NICE recommend a trial of amantadine
- other options include mindfulness training and CBT

Spasticity

- baclofen and gabapentin are first-line. Other options include diazepam, dantrolene and tizanidine
- physiotherapy is important
- cannabis and botox are undergoing evaluation

Bladder dysfunction

- may take the form of urgency, incontinence, overflow etc
- guidelines stress the importance of getting an ultrasound first to assess bladder emptying - anticholinergics may worsen symptoms in some patients
- if significant residual volume → intermittent self-catheterisation
- if no significant residual volume → anticholinergics may improve urinary frequency

Oscillopsia (visual fields appear to oscillate)

- gabapentin is first-line

External Links:

[NICE](#)

2014 Multiple Sclerosis guidelines

[Multiple Sclerosis Society](#)

Primary care guidelines

Multiple sclerosis: prognostic features

Good prognosis features

- female sex
- young age of onset (i.e. 20s or 30s)
- relapsing-remitting disease
- sensory symptoms only
- long interval between first two relapses
- complete recovery between relapses

Ways of remembering prognostic features

- the typical patient carries a better prognosis than an atypical presentation
-

Multiple system atrophy

Shy-Drager syndrome is a type of multiple system atrophy

Features

- parkinsonism
 - autonomic disturbance (atonic bladder, postural hypotension)
 - cerebellar signs
-

Myasthenia gravis

Myasthenia gravis is an autoimmune disorder resulting in insufficient functioning acetylcholine receptors. Antibodies to acetylcholine receptors are seen in 85-90% of cases*. Myasthenia is more common in women (2:1)

The key feature is muscle fatigability - muscles become progressively weaker during periods of activity and slowly improve after periods of rest:

- extraocular muscle weakness: diplopia
- proximal muscle weakness: face, neck, limb girdle
- ptosis
- dysphagia

Associations:

- thymomas in 15%
- autoimmune disorders: pernicious anaemia, autoimmune thyroid disorders, rheumatoid, SLE
- thymic hyperplasia in 50-70%

Investigations

- single fibre electromyography: high sensitivity (92-100%)
- CT thorax to exclude thymoma
- CK normal
- autoantibodies: around 85-90% of patients have antibodies to acetylcholine receptors. In the remaining patients, about 40% are positive for anti-muscle-specific tyrosine kinase antibodies
- Tensilon test: IV edrophonium reduces muscle weakness temporarily - not commonly used anymore due to the risk of cardiac arrhythmia

Management

- long-acting anticholinesterase e.g. pyridostigmine
- immunosuppression: prednisolone initially
- thymectomy.

Management of myasthenic crisis

- plasmapheresis
- intravenous immunoglobulins

*antibodies are less commonly seen in disease limited to the ocular muscles

External Links:

[Postgraduate Medical Journal](#)

Review of myasthenia gravis

Myasthenia gravis: exacerbating factors

The most common exacerbating factor is exertion resulting in fatigability, which is the hallmark feature of myasthenia gravis . Symptoms become more marked during the day

The following drugs may exacerbate myasthenia:

- penicillamine
- quinidine, procainamide
- beta-blockers
- lithium
- phenytoin
- antibiotics: gentamicin, macrolides, quinolones, tetracyclines

External Links:

[Myasthenia Gravis Association](#)

Drugs which may aggravate myasthenia gravis

[Royal College of Physicians \(Edin\)](#)

Myasthenia gravis review

Myotonic dystrophy

◆ Myotonic dystrophy (also called dystrophia myotonica) is an inherited myopathy with features developing at around 20-30 years old. It affects skeletal, cardiac and smooth muscle. There are two main types of myotonic dystrophy, DM1 and DM2.

Genetics

- autosomal dominant
- a trinucleotide repeat disorder
- DM1 is caused by a CTG repeat at the end of the DMPK (Dystrophia Myotonica-Protein Kinase) gene on chromosome 19
- DM2 is caused by a repeat expansion of the ZNF9 gene on chromosome 3

The key differences are listed in table below:

DM1	DM2
- DMPK gene on chromosome 19 - Distal weakness more prominent	- ZNF9 gene on chromosome 3 - Proximal weakness more prominent - Severe congenital form not seen

General features:

- myotonic facies (long, 'haggard' appearance)
- frontal balding
- bilateral ptosis
- cataracts
- dysarthria

Other features:

- myotonia (tonic spasm of muscle)
- weakness of arms and legs (distal initially)
- mild mental impairment
- diabetes mellitus
- testicular atrophy
- cardiac involvement: heart block, cardiomyopathy
- dysphagia.

Nerve conduction studies

Nerve conduction studies (NCS) are useful in determining between axonal and demyelinating pathology

Axonal

- normal conduction velocity
- reduced amplitude

Demyelinating

- reduced conduction velocity
- normal amplitude

External Links:

[The Bone School](#)

Nerve Conduction Studies / EMG

Neuropathic pain

Neuropathic pain may be defined as pain which arises following damage or disruption of the nervous system. It is often difficult to treat and responds poorly to standard analgesia.

Examples include:

- diabetic neuropathy
- post-herpetic neuralgia
- trigeminal neuralgia
- prolapsed intervertebral disc

NICE updated their guidance on the management of neuropathic pain in 2013:

- first-line treatment*: amitriptyline, duloxetine, gabapentin or pregabalin
- if the first-line drug treatment does not work try one of the other 3 drugs
- tramadol may be used as 'rescue therapy' for exacerbations of neuropathic pain
- topical capsaicin may be used for localised neuropathic pain (e.g. post-herpetic neuralgia)
- pain management clinics may be useful in patients with resistant problems

*please note that for some specific conditions the guidance may vary. For example carbamazepine is used first-line for trigeminal neuralgia.

External Links:

[NICE](#)

2013 Neuropathic pain guidelines

Normal pressure hydrocephalus

Normal pressure hydrocephalus is a reversible cause of dementia seen in elderly patients. It is thought to be secondary to reduced CSF absorption at the arachnoid villi. These changes may be secondary to head injury, subarachnoid haemorrhage or meningitis

A classical triad of features is seen

- urinary incontinence
- dementia and bradyphrenia
- gait abnormality (may be similar to Parkinson's disease)

Imaging

- hydrocephalus with an enlarged fourth ventricle

Management

- ventriculoperitoneal shunting

External Links:

[YouTube](#)

Typical NPH Gait

Nystagmus

Upbeat nystagmus

- cerebellar vermis lesions

Downbeat nystagmus - foramen magnum lesions

- Arnold-Chiari malformation



Image sourced from [Wikipedia](#)

Horizontal optokinetic nystagmus, a normal (physiological) form of nystagmus

Paraneoplastic syndromes affecting nervous system

Lambert-Eaton myasthenic syndrome

- associated with small cell lung cancer (also breast and ovarian)
- antibody directed against pre-synaptic voltage gated calcium channel in the peripheral nervous system
- can also occur independently as autoimmune disorder

Anti-Hu

- associated with small cell lung carcinoma and neuroblastomas
- sensory neuropathy - may be painful
- cerebellar syndrome
- encephalomyelitis

Anti-Yo

- associated with ovarian and breast cancer
- cerebellar syndrome

Anti-GAD antibody

- associated with breast, colorectal and small cell lung carcinoma
- stiff person's syndrome or diffuse hypertonia

Anti-Ri

- associated with breast and small cell lung carcinoma
 - ocular opsoclonus-myoclonus
-

Parkinson's disease: features

Parkinson's disease is a progressive neurodegenerative condition caused by degeneration of dopaminergic neurons in the substantia nigra.. This results in a classic triad of features: bradykinesia, tremor and rigidity. The symptoms of Parkinson's disease are characteristically asymmetrical.

Epidemiology

- around twice as common in men
- mean age of diagnosis is 65 years

Bradykinesia

- poverty of movement also seen, sometimes referred to as hypokinesia
- short, shuffling steps with reduced arm swinging
- difficulty in initiating movement

Tremor

- most marked at rest, 3-5 Hz
- worse when stressed or tired
- typically 'pill-rolling', i.e. in the thumb and index finger

Rigidity

- lead pipe
- cogwheel: due to superimposed tremor

Other characteristic features

- mask-like facies
- flexed posture
- micrographia
- drooling of saliva
- psychiatric features: depression is the most common feature (affects about 40%); dementia, psychosis and sleep disturbances may also occur
- impaired olfaction
- REM sleep behaviour disorder

Drug-induced parkinsonism has slightly different features to Parkinson's disease:

- motor symptoms are generally rapid onset and bilateral
- rigidity and rest tremor are uncommon

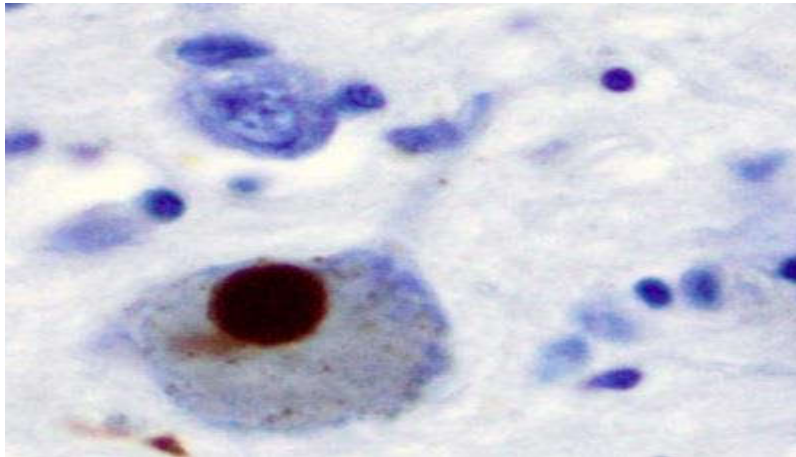


Image sourced from [Wikipedia](#)



A Lewy body (stained brown) in a brain cell of the substantia nigra in Parkinson's disease. The brown colour is positive immunohistochemistry staining for alpha-synuclein

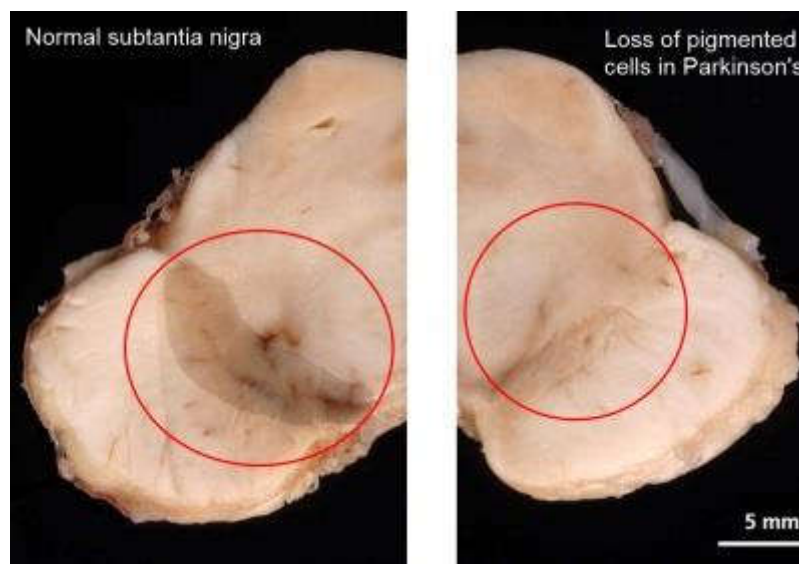


Image sourced from [Wikipedia](#)



Discoloration of the substantia nigra due to loss of pigmented nerve cells.

Parkinson's disease: management

Currently accepted practice in the management of patients with Parkinson's disease (PD) is to delay treatment until the onset of disabling symptoms and then to introduce a dopamine receptor agonist. If the patient is elderly, levodopa is sometimes used as an initial treatment.

Dopamine receptor agonists

- e.g. Bromocriptine, ropinirole, cabergoline, apomorphine
- ergot-derived dopamine receptor agonists (bromocriptine, cabergoline, pergolide*) have been associated with pulmonary, retroperitoneal and cardiac fibrosis. The Committee on Safety of Medicines advice that an echocardiogram, ESR, creatinine and chest x-ray should be obtained prior to treatment and patients should be closely monitored
- patients should be warned about the potential for dopamine receptor agonists to cause impulse control disorders and excessive daytime somnolence
- more likely than levodopa to cause hallucinations in older patients. Nasal congestion and postural hypotension are also seen in some patients

Levodopa

- usually combined with a decarboxylase inhibitor (e.g. carbidopa or benserazide) to prevent peripheral metabolism of levodopa to dopamine
- reduced effectiveness with time (usually by 2 years)
- unwanted effects: dyskinesia (involuntary writhing movements), 'on-off' effect, dry mouth, anorexia, palpitations, postural hypotension, psychosis, drowsiness
- no use in neuroleptic induced parkinsonism

MAO-B (Monoamine Oxidase-B) inhibitors

- e.g. Selegiline
- inhibits the breakdown of dopamine secreted by the dopaminergic neurons

Amantadine

- mechanism is not fully understood, probably increases dopamine release and inhibits its uptake at dopaminergic synapses
- side-effects include ataxia, slurred speech, confusion, dizziness and livedo reticularis

COMT (Catechol-O-Methyl Transferase) inhibitors:

- e.g. Entacapone, tolcapone
- COMT is an enzyme involved in the breakdown of dopamine, and hence may be used as an adjunct to levodopa therapy
- used in conjunction with levodopa in patients with established PD

Antimuscarinics:

- block cholinergic receptors
- now used more to treat drug-induced parkinsonism rather than idiopathic Parkinson's disease
- help tremor and rigidity
- e.g. procyclidine, benzotropine, trihexyphenidyl (benzhexol)

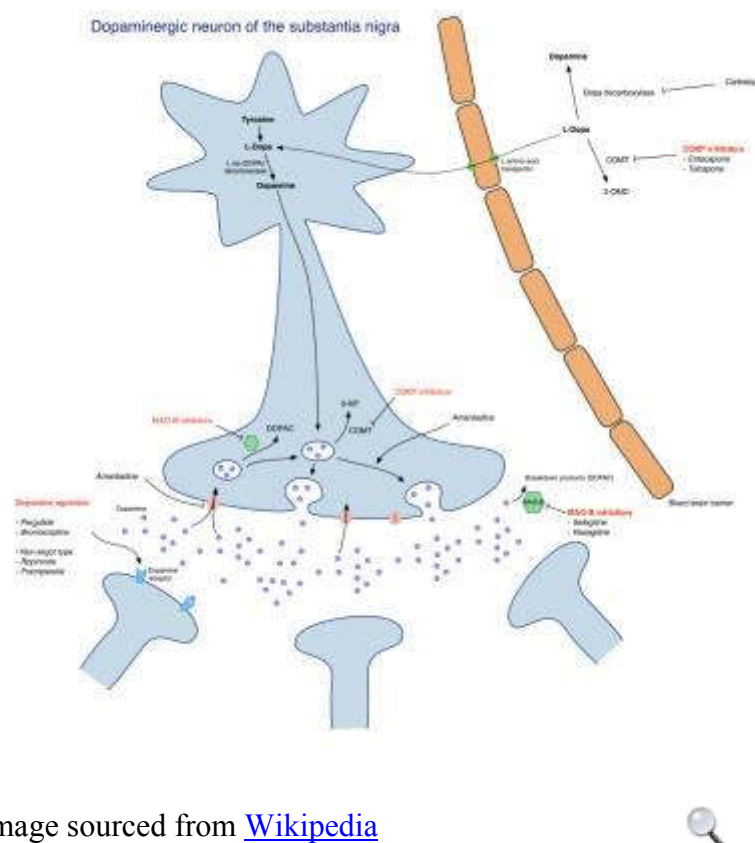


Image sourced from [Wikipedia](#)

Diagram showing the mechanism of action of Parkinson's drugs

*pergolide was withdrawn from the US market in March 2007 due to concern regarding increased incidence of valvular dysfunction

External Links:

[NICE](#)

2006 Parkinson's disease guidelines

[SIGN](#)

2010 Parkinson's disease guidelines

[Royal College of Physicians](#)

How to treat Parkinson's disease in 2013

Parkinsonism

Causes of Parkinsonism:

- Parkinson's disease
- drug-induced e.g. antipsychotics, metoclopramide - see below
- progressive supranuclear palsy
- multiple system atrophy
- Wilson's disease
- post-encephalitis
- dementia pugilistica (secondary to chronic head trauma e.g. boxing)
- toxins: carbon monoxide, MPTP

Drugs causing Parkinsonism

- phenothiazines: e.g. chlorpromazine, prochlorperazine
- butyrophenones: haloperidol, droperidol
- metoclopramide

Domperidone does not cross the blood-brain barrier and therefore does not cause extra-pyramidal side-effects.

Peripheral neuropathy

Peripheral neuropathy may be divided into conditions which predominately cause a motor or sensory loss

Predominately motor loss:

- Guillain-Barre syndrome
- porphyria
- lead poisoning
- hereditary sensorimotor neuropathies (HSMN) - Charcot-Marie-Tooth
- chronic inflammatory demyelinating polyneuropathy (CIDP)
- diphtheria

Predominately sensory loss

- diabetes
- uraemia
- leprosy
- alcoholism
- vitamin B12 deficiency
- amyloidosis

Alcoholic neuropathy

- secondary to both direct toxic effects and reduced absorption of B vitamins
- sensory symptoms typically present prior to motor symptoms

Vitamin B12 deficiency

- subacute combined degeneration of spinal cord
- dorsal column usually affected first (joint position, vibration) prior to distal paraesthesia

Peripheral neuropathy: demyelinating vs. axonal

Demyelinating pathology

- Guillain-Barre syndrome
- chronic inflammatory demyelinating polyneuropathy (CIDP)
- amiodarone
- hereditary sensorimotor neuropathies (HSMN) type I
- paraprotein neuropathy

Axonal pathology

- alcohol
- diabetes mellitus*
- vasculitis
- vitamin B12 deficiency*
- hereditary sensorimotor neuropathies (HSMN) type II

* may also cause a demyelinating picture

Pituitary apoplexy

Sudden enlargement of pituitary tumour secondary to haemorrhage or infarction

Features

- sudden onset headache similar to that seen in subarachnoid haemorrhage
 - vomiting
 - neck stiffness
 - visual field defects: classically bitemporal superior quadrantic defect
 - extraocular nerve palsies
 - features of pituitary insufficiency e.g. Hypotension secondary to hypoadrenalism
-

External Links:

[Postgraduate Medical Journal](#)

Review of endocrine emergencies

Post-lumbar puncture headache

Headache following lumbar puncture (LP) occurs in approximately one-third of patients. The pathophysiology of is unclear but may relate to a 'leak' of CSF following dural puncture. Post-LP headaches are more common in young females with a low body mass index

Typical features:

- usually develops within 24-48 hours following LP but may occur up to one week later
- may last several days
- worsens with upright position
- improves with recumbent position

Factors which may contribute to headache	Factors which do not contribute to headache
Increased needle size Direction of bevel Not replacing the stylet Increased number of LP attempts	Increased volume of CSF removed Bed rest following procedure Increased fluid intake post procedure Opening pressure of CSF Position of patient

Management

- supportive initially (analgesia, rest)
- if pain continues for more than 72 hours then specific treatment is indicated, to prevent subdural haematoma
- treatment options include: blood patch, epidural saline and intravenous caffeine

Progressive supranuclear palsy

Overview

- aka Steele-Richardson-Olszewski syndrome
- a 'Parkinson Plus' syndrome

Features

- impairment of vertical gaze (down gaze worse than up gaze - patients may complain of difficulty reading or descending stairs)
- parkinsonism
- falls
- slurring of speech
- cognitive impairment

Management

- poor response to L-dopa
-

Ptosis

Ptosis may be unilateral or bilateral

Causes of bilateral ptosis:

- myotonic dystrophy
- myasthenia gravis*
- syphilis
- congenital

Causes of unilateral ptosis, as above plus:

- third nerve palsy
- Horner's

*ptosis is much less common in Lambert-Eaton syndrome than myasthenia gravis

Restless legs syndrome

Restless legs syndrome (RLS) is a syndrome of spontaneous, continuous lower limb movements that may be associated with paraesthesia. It is extremely common, affecting between 2-10% of the general population. Males and females are equally affected and a family history may be present.

Clinical features

- uncontrollable urge to move legs (akathisia). Symptoms initially occur at night but as condition progresses may occur during the day. Symptoms are worse at rest
- paraesthesias e.g. 'crawling' or 'throbbing' sensations
- movements during sleep may be noted by the partner - periodic limb movements of sleeps (PLMS)

Causes and associations

- there is a positive family history in 50% of patients with idiopathic RLS
- iron deficiency anaemia
- uraemia
- diabetes mellitus
- pregnancy

The diagnosis is clinical although bloods to exclude iron deficiency anaemia may be appropriate.

Management

- simple measures: walking, stretching, massaging affected limbs
- treat any iron deficiency
- dopamine agonists are first-line treatment (e.g. Pramipexole, ropinirole)
- benzodiazepines
- gabapentin

Reye's syndrome

Reye's syndrome is a severe, progressive encephalopathy affecting children that is accompanied by fatty infiltration of the liver, kidneys and pancreas. The aetiology of Reye's syndrome is not fully understood although there is a known association with aspirin use and a viral cause has been postulated

The peak incidence is 2 years of age, features include:

- may be history of preceding viral illness
- encephalopathy: confusion, seizures, cerebral oedema, coma
- fatty infiltration of the liver, kidneys and pancreas
- hypoglycaemia

Management is supportive

Although the prognosis has improved over recent years there is still a mortality rate of 15-25%.

Rinne's and Weber's test

Performing both Rinne's and Weber's test allows differentiation of conductive and sensorineural deafness.

Rinne's test

- tuning fork is placed over the mastoid process until the sound is no longer heard, followed by repositioning just over external acoustic meatus
- air conduction (AC) is normally better than bone conduction (BC)
- if $BC > AC$ then conductive deafness

Weber's test

- tuning fork is placed in the middle of the forehead equidistant from the patient's ears
 - the patient is then asked which side is loudest
 - in unilateral sensorineural deafness, sound is localised to the unaffected side
 - in unilateral conductive deafness, sound is localised to the affected side
-

External Links:

[ENT SHO](#)

Guide to the ear examination

Spastic Para paresis

Spastic paraparesis describes a upper motor neuron pattern of weakness in the lower limbs

Causes:

- demyelination e.g. multiple sclerosis
- cord compression: trauma, tumour
- parasagittal meningioma
- tropical spastic paraparesis
- transverse myelitis e.g. HIV
- syringomyelia
- hereditary spastic paraplegia
- osteoarthritis of the cervical spine

Spinal cord compression

Spinal cord compression is an oncological emergency and affects up to 5% of cancer patients. Extradural compression accounts for the majority of cases, usually due to vertebral body metastases. It is more common in patients with lung, breast and prostate cancer

Features

- back pain - the earliest and most common symptom - may be worse on lying down and coughing
- lower limb weakness
- sensory changes: sensory loss and numbness
- neurological signs depend on the level of the lesion. Lesions above L1 usually result in upper motor neuron signs in the legs and a sensory level. Lesions below L1 usually cause lower motor neuron signs in the legs and perianal numbness. Tendon reflexes tend to be increased below the level of the lesion and absent at the level of the lesion.

Management

- high-dose oral dexamethasone.
- urgent oncological assessment for consideration of radiotherapy or surgery.

External Links:

[NICE](#)

2008 Metastatic spinal cord compression guidelines

Spinal cord lesions

The diagram belows shows cross-section view of the spinal cord:

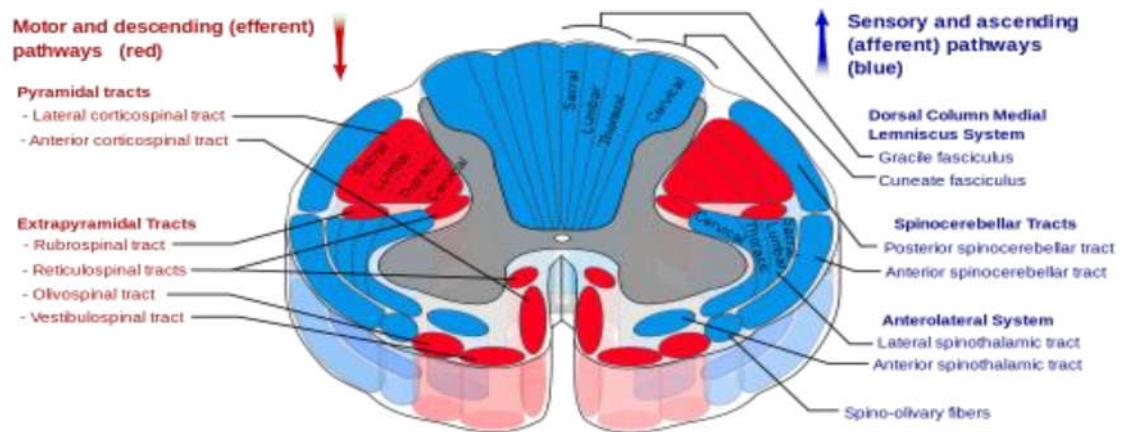


Image sourced from [Wikipedia](#)

Motor lesions

Amyotrophic lateral sclerosis (motor neuron disease):

- affects both upper (corticospinal tracts) and lower motor neurons
- results in a combination of upper and lower motor neuron signs

Poliomyelitis:

- affects anterior horns resulting in lower motor neuron signs.

Combined motor and sensory lesions

Disorder	Tracts affected	Clinical notes
Brown-Sequard syndrome (spinal cord hemisection)	1. Lateral corticospinal tract 2. Dorsal columns 3. Lateral spinothalamic tract	1. Ipsilateral spastic paresis below lesion 2. Ipsilateral loss of proprioception and vibration sensation 3. Contralateral loss of pain and temperature sensation
Subacute combined degeneration of the spinal cord (vitamin B12 & E deficiency)	1. Lateral corticospinal tracts 2. Dorsal columns 3. Spinocerebellar tracts	1. Bilateral spastic paresis 2. Bilateral loss of proprioception and vibration sensation 3. Bilateral limb ataxia
Friedrich's ataxia	Same as subacute combined degeneration of the spinal cord (see above)	Same as subacute combined degeneration of the spinal cord (see above) In addition cerebellar ataxia → other features e.g. intention tremor
Anterior spinal artery occlusion	1. Lateral corticospinal tracts 2. Lateral spinothalamic tracts	1. Bilateral spastic paresis 2. Bilateral loss of pain and temperature sensation
Syringomyelia	1. Ventral horns 2. Lateral spinothalamic tract	1. Flacid paresis (typically affecting the intrinsic hand muscles) 2. Loss of pain and temperature sensation
Multiple sclerosis	Asymmetrical, varying spinal tracts involved	Combination of motor, sensory and ataxia symptoms

Sensory lesions

Disorder	Tracts affected	Clinical notes
Neurosypilis (tabes dorsalis)	1. Dorsal columns	1. Loss of proprioception and vibration sensation

Stroke by anatomy

Site of the lesion	Associated effects
Anterior cerebral artery	Contralateral hemiparesis and sensory loss, lower extremity > upper
Middle cerebral artery	Contralateral hemiparesis and sensory loss, upper extremity > lower Contralateral homonymous hemianopia Aphasia
Posterior cerebral artery	Contralateral homonymous hemianopia with macular sparing Visual agnosia
Weber's syndrome (branches of the posterior cerebral artery that supply the midbrain)	Ipsilateral CN III palsy Contralateral weakness of upper and lower extremity
Posterior inferior cerebellar artery (lateral medullary syndrome, Wallenberg syndrome)	Ipsilateral: facial pain and temperature loss Contralateral: limb/torso pain and temperature loss Ataxia, nystagmus
Anterior inferior cerebellar artery (lateral pontine syndrome)	Symptoms are similar to Wallenberg's (see above), but: Ipsilateral: facial paralysis and deafness
Retinal/ophthalmic artery	Amaurosis fugax
Basilar artery	'Locked-in' syndrome

Lacunar strokes

- present with either isolated hemiparesis, hemisensory loss or hemiparesis with limb ataxia.
- strong association with hypertension.
- common sites include the basal ganglia, thalamus and internal capsule.

Stroke: management

The Royal College of Physicians (RCP) published guidelines on the diagnosis and management of patients following a stroke in 2004. NICE also issued stroke guidelines in 2008, although they modified their guidance with respect to antiplatelet therapy in 2010.

Selected points relating to the management of acute stroke include:

- blood glucose, hydration, oxygen saturation and temperature should be maintained within normal limits
- blood pressure should not be lowered in the acute phase unless there are complications e.g. Hypertensive encephalopathy*
- aspirin 300mg orally or rectally should be given as soon as possible if a haemorrhagic stroke has been excluded
- with regards to atrial fibrillation, the RCP state: 'anticoagulants should not be started until brain imaging has excluded haemorrhage, and usually not until 14 days have passed from the onset of an ischaemic stroke'
- if the cholesterol is > 3.5 mmol/l patients should be commenced on a statin. Many physicians will delay treatment until after at least 48 hours due to the risk of haemorrhagic transformation

Thrombolysis

Thrombolysis should only be given if:

- it is administered within 4.5 hours of onset of stroke symptoms (unless as part of a clinical trial)
- haemorrhage has been definitively excluded (i.e. Imaging has been performed)

♦Alteplase is currently recommended by NICE.

Contraindications to thrombolysis:

Absolute	Relative
- Previous intracranial haemorrhage - Seizure at onset of stroke - Intracranial neoplasm - Suspected subarachnoid haemorrhage - Stroke or traumatic brain injury in preceding 3 months - Lumbar puncture in preceding 7 days - Gastrointestinal haemorrhage in preceding 3 weeks - Active bleeding - Pregnancy - Oesophageal varices - Uncontrolled hypertension $>200/120$mmHg	- Concurrent anticoagulation (INR >1.7) - Haemorrhagic diathesis - Active diabetic haemorrhagic retinopathy - Suspected intracardiac thrombus - Major surgery / trauma in preceding 2 weeks

Secondary prevention

NICE also published a technology appraisal in 2010 on the use of clopidogrel and dipyridamole.

Recommendations from NICE include:

- clopidogrel is now recommended by NICE ahead of combination use of aspirin plus modified release (MR) dipyridamole in people who have had an ischaemic stroke
- aspirin plus MR dipyridamole is now recommended after an ischaemic stroke only if clopidogrel is contraindicated or not tolerated, but treatment is no longer limited to 2 years' duration
- MR dipyridamole alone is recommended after an ischaemic stroke only if aspirin or clopidogrel are contraindicated or not tolerated, again with no limit on duration of treatment

With regards to carotid artery endarterectomy:

- recommend if patient has suffered stroke or TIA in the carotid territory and are not severely disabled
- should only be considered if carotid stenosis > 70% according ECST** criteria or > 50% according to NASCET*** criteria

*the 2009 Controlling hypertension and hypotension immediately post-stroke (CHHIPS) trial may change thinking on this but guidelines have yet to change to reflect this

**European Carotid Surgery Trialists' Collaborative Group

***North American Symptomatic Carotid Endarterectomy Trial

External Links

[NICE](#)

2008 Stroke guidelines

[SIGN](#)

2008 Stroke guidelines

[RCP](#)

Stroke guidelines

[NICE](#)

2010 Clopidogrel and dipyridamole guidelines

[Age and Ageing](#)

Interesting article on managing blood pressure during acute stroke

[Clinical Knowledge Summaries](#)

Stroke and TIA guidelines

Stroke: types

The **Oxford Stroke Classification** (also known as the Bamford Classification) classifies strokes based on the initial symptoms. A summary is as follows:

The following criteria should be assessed:

- 1. unilateral hemiparesis and/or hemisensory loss of the face, arm & leg
- 2. homonymous hemianopia
- 3. higher cognitive dysfunction e.g. dysphasia

Total anterior circulation infarcts (TACI, c. 15%)

- involves middle and anterior cerebral arteries
- all 3 of the above criteria are present

Partial anterior circulation infarcts (PACI, c. 25%):

- involves smaller arteries of anterior circulation e.g. upper or lower division of middle cerebral artery
- 2 of the above criteria are present

Lacunar infarcts (LACI, c. 25%)

- involves perforating arteries around the internal capsule, thalamus and basal ganglia
- presents with 1 of the following:
 - 1. unilateral weakness (and/or sensory deficit) of face and arm, arm and leg or all three.
 - 2. pure sensory stroke.
 - 3. ataxic hemiparesis

Posterior circulation infarcts (POCI, c. 25%):

- involves vertebrobasilar arteries
- presents with 1 of the following:
 - 1. cerebellar or brainstem syndromes
 - 2. loss of consciousness
 - 3. isolated homonymous hemianopia.

Other recognized patterns of stroke:

Lateral medullary syndrome (posterior inferior cerebellar artery)

- aka Wallenberg's syndrome
- ipsilateral: ataxia, nystagmus, dysphagia, facial numbness, cranial nerve palsy e.g. Horner's
- contralateral: limb sensory loss

Weber's syndrome

- ipsilateral III palsy
- contralateral weakness

External Links

[YouTube](#)

Bamford Classification

Subarachnoid hemorrhage

Causes:

- 85% are due to rupture of berry aneurysms (conditions associated with berry aneurysms include adult polycystic kidney disease, Ehlers-Danlos syndrome and coarctation of the aorta)
- AV malformations
- trauma
- tumours

Investigations:

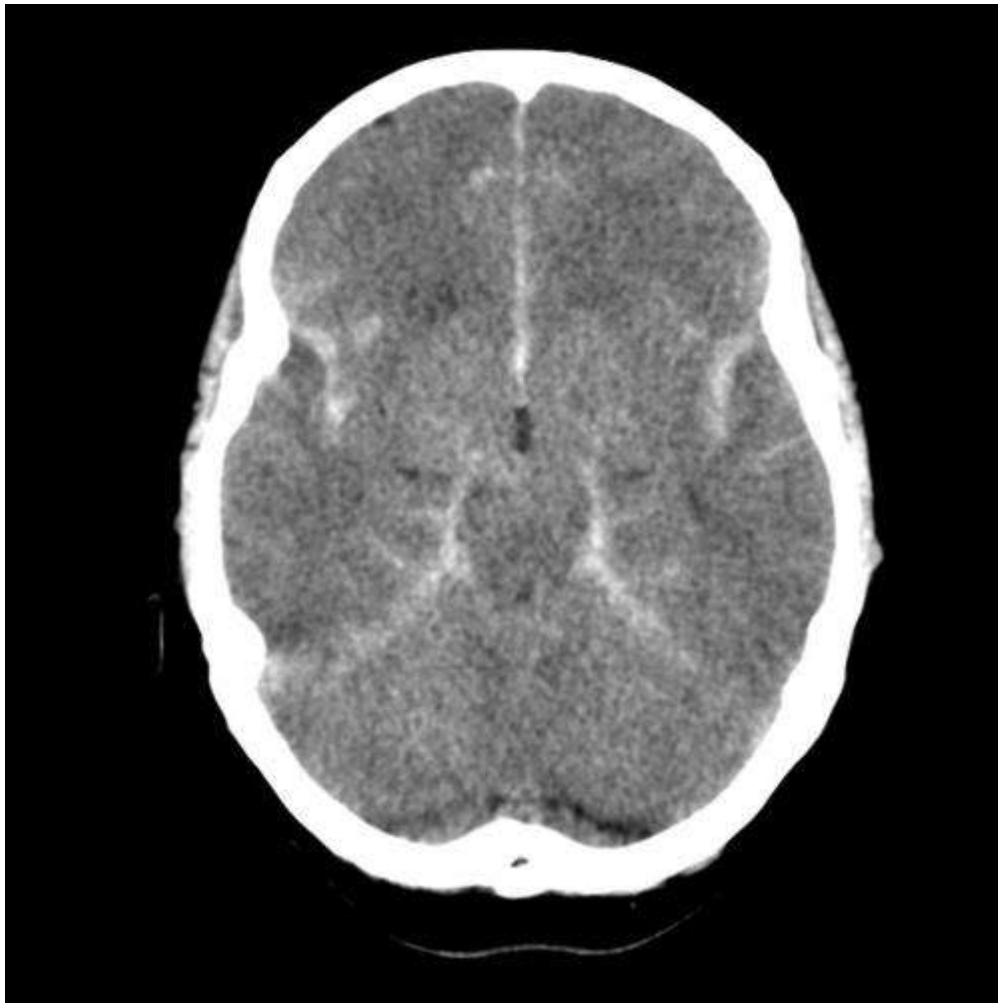
- CT: negative in 5%
- lumbar puncture: done after 12 hrs (allowing time for xanthochromia to develop)

Complications:

- rebleeding (in 30%)
- obstructive hydrocephalus (due to blood in ventricles)
- vasospasm leading to cerebral ischaemia

Management

- neurosurgical opinion: no clear evidence over early surgical intervention against delayed intervention
- post-operative nimodipine (e.g. 60mg / 4 hrly, if BP allows) has been shown to reduce the severity of neurological deficits but doesn't reduce rebleeding*



© Image used on license from [Radiopaedia](#)

CT image shows diffuse subarachnoid haemorrhage in all basal cisterns, bilateral sylvian fissures and the inter-hemispheric fissure. This case demonstrates the typical distribution that takes the blood into the subarachnoid space in a subarachnoid hemorrhage.

*the way nimodipine works in subarachnoid haemorrhage is not fully understood. It has been previously postulated that it reduces cerebral vasospasm (hence maintaining cerebral perfusion) but this has not been demonstrated in studies

External Link

[SIGN](#)

2008 Diagnosis and management of headache in adults

Subdural hemorrhage

Basics

- most commonly secondary to trauma e.g. old person/alcohol falling over
- initial injury may be minor and is often forgotten
- caused by bleeding from damaged bridging veins between cortex and venous sinuses

Features

- headache
- classically fluctuating conscious level
- raised ICP

Treatment

- needs neurosurgical review ? burr hole

External Links

[Postgraduate Medical Journal](#)

Chronic subdural haematoma

Syringomyelia

Overview

- development of cavity (syrinx) within the spinal cord
- if extends into medulla then termed syringobulbia
- strongly associated with the Arnold-Chiari malformation

Features

- maybe asymmetrical initially
- slowly progressives, possibly over years
- motor: wasting and weakness of arms
- sensory: spinothalamic sensory loss (pain and temperature)
- loss of reflexes, bilateral upgoing plantars
- also seen: Horner's syndrome

Tinnitus

Causes of tinnitus include:

Meniere's disease	Associated with hearing loss, vertigo, tinnitus and sensation of fullness or pressure in one or both ears
Otosclerosis	Onset is usually at 20-40 years Conductive deafness Tinnitus Normal tympanic membrane* Positive family history
Acoustic neuroma	Hearing loss, vertigo, tinnitus Absent corneal reflex is important sign Associated with neurofibromatosis type 2
Hearing loss	Causes include excessive loud noise and presbycusis
Drugs	Aspirin Aminoglycosides Loop diuretics Quinine

Other causes include:

- impacted ear wax
- chronic suppurative otitis media

*10% of patients may have a 'flamingo tinge', caused by hyperaemia

Transient global amnesia

Overview

- presents with transient loss of memory function
- patients may appear anxious and repeatedly ask the same question
- patients have no recall of events after the attack
- etiology is unknown, thought to be due to transient ischaemia to the thalamus (in particular the amygdala and hippocampus)

Transient ischemic attack

NICE issued updated guidelines relating to stroke and transient ischaemic attack (TIA) in 2008. **They advocated the use of the ABCD2 prognostic score for risk stratifying patients who've had a suspected TIA:**

	Criteria	Points
A	Age $\geq$ 60 years	1
B	Blood pressure $\geq$ 140/90 mmHg	1
C	Clinical features	
	- Unilateral weakness	2
	- Speech disturbance, no weakness	1
D	Duration of symptoms	
	- > 60 minutes	2
	- 10-59 minutes	1
	Patient has diabetes	1

This gives a total score ranging from 0 to 7. **People who have had a suspected TIA who are at a higher risk of stroke (that is, with an ABCD2 score of 4 or above) should have:**

- aspirin (300 mg daily) started immediately.
- specialist assessment and investigation within 24 hours of onset of symptoms.
- measures for secondary prevention introduced as soon as the diagnosis is confirmed, including discussion of individual risk factors.

If the ABCD2 risk score is 3 or below:

- specialist assessment within 1 week of symptom onset, including decision on brain imaging
- if vascular territory or pathology is uncertain, refer for brain imaging

People with crescendo TIAs (two or more episodes in a week) should be treated as being at high risk of stroke, even though they may have an ABCD2 score of 3 or below.

Antithrombotic therapy:

- clopidogrel is recommended first-line (as for patients who've had a stroke)
- aspirin + dipyridamole should be given to patients who cannot tolerate clopidogrel
- these recommendations follow the 2012 Royal College of Physicians National clinical guideline for stroke. Please see the link for more details (section 5.5)
- these guidelines may change following the CHANCE study (NEJM 2013;369:11). This study looked at giving high-risk TIA patients aspirin + clopidogrel for the first 90 days compared to aspirin alone. 11.7% of aspirin only patients had a stroke over 90 days compared to 8.2% of dual antiplatelet patients

With regards to carotid artery endarterectomy:

- recommend if patient has suffered stroke or TIA in the carotid territory and are not severely disabled
- should only be considered if carotid stenosis > 70% according ECST* criteria or > 50% according to NASCET** criteria

*European Carotid Surgery Trialists' Collaborative Group

**North American Symptomatic Carotid Endarterectomy Trial

External Links

[NICE](#)

2008 Stroke: Diagnosis and initial management of acute stroke and transient ischaemic attack (TIA)

[Royal College of Physicians](#)

2016 National clinical guideline for stroke

Trigeminal neuralgia

Trigeminal neuralgia is a pain syndrome characterised by severe unilateral pain. The vast majority of cases are idiopathic but compression of the trigeminal roots by tumours or vascular problems may occur.

The International Headache Society defines trigeminal neuralgia as:

- a unilateral disorder characterised by brief electric shock-like pains, abrupt in onset and termination, limited to one or more divisions of the trigeminal nerve
- the pain is commonly evoked by light touch, including washing, shaving, smoking, talking, and brushing the teeth (trigger factors), and frequently occurs spontaneously
- small areas in the nasolabial fold or chin may be particularly susceptible to the precipitation of pain (trigger areas)
- the pains usually remit for variable periods

Management:

- carbamazepine is first-line
- failure to respond to treatment or atypical features (e.g. < 50 years old) should prompt referral to neurology

External Links

[Clinical Knowledge Summaries](#)

Trigeminal Neuralgia guidelines

Triptans

Triptans are specific 5-HT₁ agonists used in the acute treatment of migraine. They are generally used first-line in combination therapy with an NSAID or paracetamol.

Prescribing points:

- should be taken as soon as possible after the onset of headache, rather than at onset of aura
- oral, orodispersible, nasal spray and subcutaneous injections are available

Adverse effects:

- 'triptan sensations' - tingling, heat, tightness (e.g. throat and chest), heaviness, pressure

Contraindications:

- patients with a history of, or significant risk factors for, ischaemic heart disease or cerebrovascular disease

External Links

[Clinical Knowledge Summaries](#)

Migraine guidelines

Tuberous sclerosis

Tuberous sclerosis (TS) is a genetic condition of autosomal dominant inheritance. Like neurofibromatosis, the majority of features seen in TS are neuro-cutaneous

Cutaneous features:

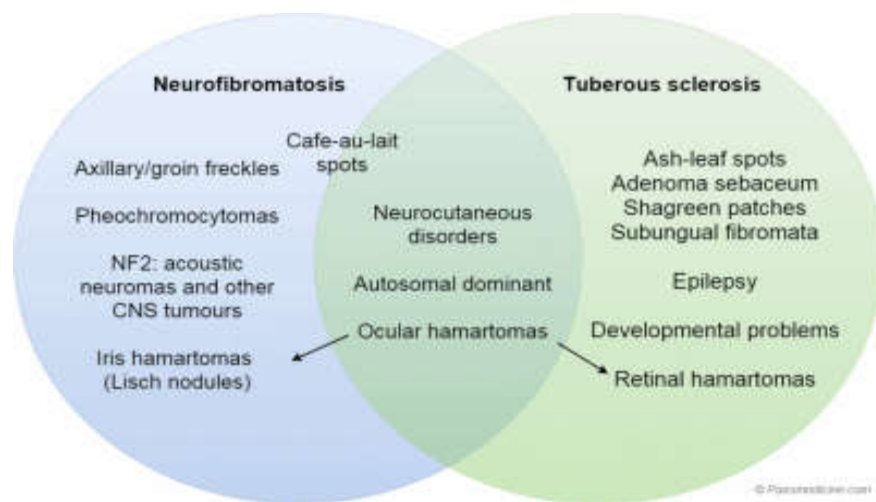
- depigmented 'ash-leaf' spots which fluoresce under UV light
- roughened patches of skin over lumbar spine (Shagreen patches)
- adenoma sebaceum (angiofibromas): butterfly distribution over nose
- fibromata beneath nails (subungual fibromata)
- café-au-lait spots* may be seen

Neurological features

- developmental delay
- epilepsy (infantile spasms or partial)
- intellectual impairment

Also

- retinal hamartomas: dense white areas on retina (phakomata)
- rhabdomyomas of the heart
- gliomatous changes can occur in the brain lesions
- polycystic kidneys, renal angiomyolipomata
- lymphangiomyomatosis: multiple lung cysts



Comparison of neurofibromatosis and tuberous sclerosis. Note that whilst they are both autosomal dominant neurocutaneous disorders there is little overlap otherwise

*these of course are more commonly associated with neurofibromatosis. However a 1998 study of 106 children with TS found café-au-lait spots in 28% of patients

External Links

[Patient.info](#)

Tuberous sclerosis review

Vigabatrin

Key points

- 40% of patients develop visual field defects, which may be irreversible
- visual fields should be checked every 6 months

Visual field defects

The main points for the exam are:

- left homonymous hemianopia means visual field defect to the left, i.e. Lesion of right optic tract
- homonymous quadrantanopias: PITS (Parietal-Inferior, Temporal-Superior)
- incongruous defects = optic tract lesion; congruous defects = optic radiation lesion or occipital cortex

A congruous defect simply means complete or symmetrical visual field loss and conversely an incongruous defect is incomplete or asymmetric. Please see the link for an excellent diagram.

Homonymous hemianopia:

- incongruous defects: lesion of optic tract
- congruous defects: lesion of optic radiation or occipital cortex
- macula sparing: lesion of occipital cortex

Homonymous quadrantanopias*:

- superior: lesion of temporal lobe
- inferior: lesion of parietal lobe
- mnemonic = PITS (Parietal-Inferior, Temporal-Superior)

Bitemporal hemianopia:

- lesion of optic chiasm
- upper quadrant defect > lower quadrant defect = inferior chiasmal compression, commonly a pituitary tumour
- lower quadrant defect > upper quadrant defect = superior chiasmal compression, commonly a craniopharyngioma

*this is very much the 'exam answer'. Actual studies suggest that the majority of quadrantanopias are caused by occipital lobe lesions. Please see the following link for more details.

External Links

[Patient.info](#)

Visual field defects

[PubMed](#)

The localizing value of a quadrantanopia

Von Hippel-Lindau syndrome

Von Hippel-Lindau (VHL) syndrome is an autosomal dominant condition predisposing to neoplasia. It is due to an abnormality in the VHL gene located on short arm of chromosome 3

Features

- cerebellar haemangiomas
- retinal haemangiomas: vitreous haemorrhage
- renal cysts (premalignant)
- pheochromocytoma
- extra-renal cysts: epididymal, pancreatic, hepatic
- endolymphatic sac tumours



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CT scan showing a cerebellar haemangioma in a patient with Von Hippel-Lindau syndrome.



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MRI showing renal cysts in patient with known Von Hippel-Lindau syndrome.
